ANNUAL REPORT 2019

Advancing treatment in neurodegenerative orphan diseases

Harvey Charman
Aged 3, suffers from Niemann-Pick disease Type C (NPC), having fun on the Children's Activity Program at the NPUK Annual Family Conference

ORPHAZYME

TABLE OF CONTENTS

AT A GLANCE

Pioneering the Heat-Shock Protein response for neurodegenerative orphan diseases

3

ORPHAN DRUG DESIGNATIONS

2

FAST TRACK DESIGNATIONS

1

BREAKTHROUGH THERAPY DESIGNATION

ARIMOCLOMOL

First-in-class Heat-Shock Protein (HSP) amplifier

Readily crosses blood-brain barrier

Easy oral, nasogastric administration

Favorable safety profile

3

PIVOTAL TRIALS

Niemann-Pick disease Type C (NPC)

Amyotrophic Lateral Sclerosis (ALS)

Sporadic Inclusion Body Myositis (sIBM)

1

PHASE 2 TRIAL

in Gaucher disease

GET TO APPROVAL, GET TO PATIENTS

Preparing to submit arimoclomol for approval in US and EU for NPC

Copenhagen, Denmark
HQ, Listed on Nasdaq Copenhagen (ORPHA.CO)
Boston, US
US subsidiary
Zug, Switzerland
CH subsidiary

2019 ACHIEVEMENTS

JANUARY

Orphazyme reports positive results from phase 2/3 trial in NPC

APRIL

Orphazyme's phase 2/3 trial in sIBM fully enrolled

JUNE

Orphazyme to prepare for filing of arimoclomol in Europe for NPC

JULY

Orphazyme announces appointment of Kim Stratton as Chief Executive Officer

Orphazyme completes enrollment in phase 3 trial evaluating arimoclomol in ALS

Orphazyme to prepare for filing of arimoclomol in US for NPC

NOVEMBER

Orphazyme receives Breakthrough Therapy Designation for arimoclomol in NPC from FDA

DECEMBER

Orphazyme receives Fast-Track designation for arimoclomol in sIBM

Priority	✓	Targeted milestone
ALS	✓	Complete enrollment in H2 2019
sIBM	✓	Complete enrollment in H1 2019
NPC	✓	Regulatory feedback mid-2019
Gaucher disease		Phase 2 results in H2 2019
NME program	✓	Preclinical studies with NMEs in protein-misfolding diseases

POISED FOR COMMERCIAL STAGE

This year marked Orphazyme's 10th anniversary. Having served as a member of the Board of Directors for many years, it has been particularly rewarding to witness the Company's evolution from humble beginnings to a thriving enterprise with a clear mission to bring innovative new therapies to patients suffering from rare diseases with no or limited treatment options available.

Orphazyme's lead investigational candidate, arimoclomol, is now at a pivotal stage in its development as the Company is poised to become a commercial-stage biopharmaceutical company with a proprietary portfolio within rare diseases. Orphazyme is preparing regulatory submissions for arimoclomol in the US and EU this year, and expects approvals of arimoclomol in NPC in 2021. Due to the Company's fast development towards bringing its first product to the market, it was a natural step to bring in Kim Stratton as Chief Executive Officer. Kim brings more than 25 years' industry experience, with specific expertise in managing commercial organizations operating within rare diseases. She is a proven leader who has successfully built and led high-performance teams to deliver sustainable business results, and Orphazyme is already now benefitting from her leadership since she started as CEO in October.

I would also like to take a moment to extend my deepest gratitude to Anders Hinsby, our outgoing CEO, who is a Co-Founder of Orphazyme. Anders has been instrumental in the successful building up of the company from early-stage R&D to pre-commercial, and I am very grateful for his important contribution to Orphazyme and wish him all the best for the future.

It is very satisfactory to record that Orphazyme has consistently and successfully delivered a number of significant milestones during 2019. In NPC, the company reported positive results from its phase 2/3 trial, gained positive feedback from FDA and EMA on the paths to approval and was granted Breakthrough Therapy Designation from the FDA for arimoclomol. Additionally, Orphazyme completed enrollment in its sIBM, ALS, and Gaucher disease trials and was awarded Fast Track designation from the FDA for arimoclomol in sIBM. Collectively, these development milestones demonstrate a company that is executing exceptionally well, advancing arimoclomol closer to the market and patients, and, importantly, paving the way to expand arimoclomol's potential beyond NPC into other indications.

It is an honor to serve as Chairman of Orphazyme and I am continuously looking forward to overseeing its further evolution. Given the progress outlined above, I am confident that 2020 will mark another year of reaching important milestones. On behalf of the Board of Directors, I would like to thank our shareholders for their continued belief in the company, without which these advances in medicine would not be possible.

Best Regards,

Georges Gemayel
Chairman of the
Board of Directors

LETTER FROM THE CEO

FOCUS ON GETTING TO PATIENTS

At Orphazyme, we are pioneering the Heat-Shock Protein response for patients with neurodegenerative orphan diseases – this is our purpose. We have made significant progress in bringing our innovative therapy to rare disease patients in need and we are now close to launch in our first indication, Niemann-Pick disease Type C (NPC).

2019 was an exceptional year for Orphazyme: In Niemann-Pick disease Type C (NPC), we reported positive data from our phase 2/3 trial, received Breakthrough Therapy Designation from the FDA and, very early in 2020, reported data from our 12-month open-label extension trial and launched our Early Access Program in the US. In ALS, we completed recruitment of our phase 3 trial sooner than expected, thanks to the global ALS community, patients, and caregivers, and in sporadic Inclusion Body Myositis (sIBM) we completed enrollment of our phase 2/3 trial and received Fast-Track designation from the FDA. 2019 was also the year in which we celebrated our 10-year anniversary as a company. All milestones that strongly underline the potential of our compound.

Early in the year we reported positive data from our pivotal phase 2/3 trial in patients with NPC. Treatment with arimoclomol adjunct to routine clinical care resulted in a reduction in disease progression when compared to the placebo group after 12 months. Statistically significant reductions in disease progression were observed in two pre-specified subgroups; patients aged ≥4 years and routine clinical care with miglustat and, as subsequently reported, in a post-hoc subgroup which removed patients with double functional null mutations. Arimoclomol was well-tolerated with a similar incidence of adverse events as with the placebo groups.

> In 2019, Orpahzyme celebrated its 10-year anniversary. We are now on the cusp of getting a new therapy to patients

Our development team continued to explore the potential of arimoclomol in additional indications and during the year completed enrollment in two pivotal trials: one for Amyotrophic Lateral Sclerosis (ALS), a fatal, rapidly progressive, devastating neurodegenerative disease and in sporadic Inclusion Body Myositis (sIBM): A relentlessly progressive debilitating muscle-wasting neuromuscular disease that leads to loss of mobility and independence. Both of these trials are set to report top-line data in H1 2021.

More work remains to be done for rare disease patients. Patients with Gaucher Type I and III suffer from severe neurological symptoms and currently have no access to treatment targeting the central nervous system (CNS) symptoms. We expect results from our phase 2 Gaucher disease trial in H1 2020.

In 2020, we will be preparing for filing and launch of arimoclomol in NPC. We are on track to submit applications in the US in H1 2020 and in EU in H2 2020. We are in the process of building our go-to-market capabilities and have brought in strong talent with substantial rare disease expertise to support Orphazyme in our efforts to bring arimoclomol to many patients around the world so that we can profoundly impact the lives of patients and families living with orphan diseases.

We are committed to bringing treatment to our NPC patients as soon as possible and recently launched our Early Access Program in the US. In 2020, we aim to expand the Early Access Programs to more countries around the world.

In early 2020 we significantly strengthened our balance sheet, raising DKK 745 million (USD 110 million) in a directed issue and private placement. This funding provides a significant cash runway for Orphazyme as we progress our lead product candidate arimoclomol towards market and is expected to cover our ambitious clinical development plans well into 2021, including completion of trials in sIBM and ALS and preparations for filings in EU and potentially the US in these indications.

Since joining the company as Chief Executive Officer in October last year, I have been impressed by the dedication and commitment of our team. Our purpose is inspired by the potential to address the major unmet medical needs for our patients and their families. Now, we are within reach of our goal of bringing our innovative medicine arimoclomol to our first group of patients with NPC. I would like to take this opportunity to extend my thanks to all our employees. It is their hard work and dedication that will enable us to deliver on our promises. I also want to thank our patients, patient groups, physicians, and all of our key stakeholders for their continued trust and support.

Yours Sincerely,

Kim Stratton
Chief Executive Officer

KEY FIGURES

(TDKK)	2019	2018	2017	2016	2015[1]
Statement of profit and loss and other comprehensive income					
Research and development expenses	(285,413)	(196,525)	(99,048)	(55,817)	(25,478)
General and administrative expenses	(50,541)	(35,127)	(31,994)	(7,703)	(4,044)
Operating loss	**(335,954)**	**(231,652)**	**(131,042)**	**(63,520)**	**(29,522)**
Net financial items	(7,043)	(3,448)	(662)	85	40
Loss before tax	**(342,997)**	**(235,100)**	**(131,704)**	**(63,435)**	**(29,482)**
Income tax benefit	5,500	5,500	5,500	5,500	2,750
Net loss for the period	**(337,497)**	**(229,600)**	**(126,204)**	**(57,935)**	**(26,732)**
Total comprehensive loss	**(337,430)**	**(229,558)**	**(126,204)**	**(57,935)**	**(26,732)**
Loss per share, basic (DKK)	(16.87)	(11.49)	(10.43)	(5.89)	(2.75)
Statement of financial position					
Licenses	10,539	10,744	9,853	-	-
Right-of-use assets	13,903	-	-	-	-
Property, plant, and equipment	3,685	1,940	1,851	1,225	1,512
Total non-current assets	32,529	17,965	14,864	4,047	4,448
Cash	123,588	394,706	631,735	14,349	68,014
Other current assets	19,137	28,678	16,218	13,545	12,490
Total assets	180,754	441,349	662,817	31,941	84,952
Share capital	19,984	19,939	19,928	3,361	3,346
Total equity	52,969	388,249	615,702	17,509	74,143
Non-current borrowings	51,606	-	-	-	-
Non-current lease liabilities	9,813	-	-	-	-
Total current liabilities	65,988	52,995	47,115	14,432	10,809

(TDKK)	2019	2018	2017	2016[1]	2015
Cash flow statement					
Cash flow from operating activities	(326,818)	(234,764)	(95,426)	(54,724)	(21,372)
Cash flow from investing activities	(3,285)	(2,346)	(1,491)	(238)	(25)
Cash flow from financing activities	58,939	-	714,303	1,300	11,250
Other					
Share price (DKK)[3]	72.40	43.35	76.00	-	-
Total outstanding shares	19,984,799	19,939,564	19,928,184	3,360,541	3,345,755
Market capitalization (MDKK)	1,446.9	864.4	1,514.5	-	-
Equity ratio[5]	29%	88.0%	92.9%	54.8%	87.3%
Equity per share (DKK)[6]	2.65	19.47	30.90	5.21	22.16
Average number of employees	74	46	26	17	13
Number of employees at the end of the year	86	57	34	21	15

The comparatives figures for 2018-2015 have not been restated following the adoption of IFRS 16 *Leases* on January 1, 2019 by use of the modified retrospective approach.

[1] 2015 covers the period July 1, 2015 to December 31, 2015 as the Company changed its financial year to correspond to the calendar year
[3] There is no official share price for the reporting periods prior to 2017 since the Company only went public in 2017
[4] Market capitalization is calculated as the share price multiplied with the total outstanding shares as of the balance sheet date
[5] Equity ratio is calculated as the equity divided by the total assets as of the balance sheet date
[6] Equity per share is calculated as the total equity divided by the total outstanding shares as of the balance sheet date

2020 OUTLOOK

(MDKK)	2020 guidance	2019 actual result	2019 guidance
Operating loss	(500) - (550)	(336)	(315) - (345)
Cash position at year-end	>300	124	>110

OPERATING RESULT

The operating loss of DKK 336 million for the year ended December 31, 2019 is within the expected operating loss range of DKK 315–345 million. We anticipate that our 2020 operating loss will be in the range of DKK 500–550 million. This range reflects the inherent uncertainty related to the timing of patient enrollment and related operational uncertainties. The cost increase is driven by our preparation to submit a New Drug Application/Marketing Authorization Application for arimoclomol for NPC, advancement of arimoclomol for sIBM and ALS, production of arimoclomol, and an increase in employees to support our expanding clinical and operational activities.

CASH POSITION

At December 31, 2020 we anticipate a cash position greater than DKK 300 million compared to DKK 124 million as of December 31, 2019.

RISKS AND ASSUMPTIONS

In addition to factors already mentioned, the estimates above are subject to change due to numerous reasons, including but not limited to the timing and extent of commercial preparation and development activities.

For the financial year ending December 31, 2020, Orphazyme expects to ramp up costs associated with filing and launch activities and to continue to incur significant costs associated with clinical trials, which are fully recruited.

The forecasting of costs associated with clinical trials relating to activities performed by Clinical Research Organizations (CROs) and other external vendors requires management to exercise significant estimates with regard to the timing and accounting for these costs. The diverse nature of the services being provided by CROs and other arrangements, the different compensation arrangements that exist for each type of service, and the limitations in respect of information related to certain clinical activities add complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. The filing activities for arimoclomol for NPC will be influenced by continuous regulatory feedback from the US Food & Drug Administration (FDA) and the European Medicines Agency (EMA) and the ongoing preclinical activities to be completed during 2020. Launch activities will significantly ramp up during 2020 and be considerably influenced by the roll-out of our commercial strategy and potential market entries.

The outlook for the financial year ending December 31, 2020 takes into consideration activities planned for 2020 as described on page 12, Product Pipeline.

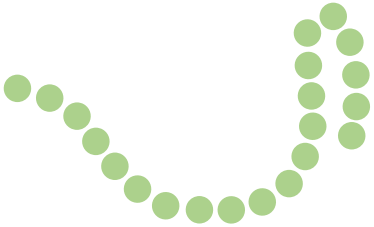
Disclaimer

This annual report contains forward-looking statements. The words “believe”, “expect”, “anticipate”, “intend”, “plan”, and similar expressions identify forward-looking statements, including in respect of the financial and business outlook described above. Actual results or performance may differ materially from any future results or performance expressed or implied by such statements. The important factors that could cause our actual results or performance to differ materially include, among others, risks associated with product discovery and development, uncertainties related to the outcome and conduct of clinical trials, including unforeseen safety issues, uncertainties related to product manufacturing, and other factors. For a further discussion of these risks, please refer to the section Risk Management on pages 21-22 in this annual report. Orphazyme A/S does not undertake any obligation to update or revise forward-looking statements in this annual report nor to confirm such statements in relation to actual results, unless required by law.

PRIORITIES 2020 AND BEYOND

Priority	Targeted Milestone	Estimated Timing
Get to approval in NPC	Submit New Drug Application US	H1 2020
	Submit Marketing Authorization Application in EU	H2 2020
Get to patients in NPC	US Early Access Program roll-out	2020
	Preparations for US launch	2020
	Lean go-to market roll out US	2020
	Lean go-to market roll out EU/ROW	H2 2020 - 2021
Expand beyond NPC	Phase 2 results Gaucher disease	H1 2020
	Phase 2/3 results sIBM	H1 2021
	Phase 3 results ALS	H1 2021

We are within reach of our goal of bringing our innovative medicine arimoclomol to our first group of patients with NPC



Pioneering the Heat-Shock Protein response for neurodegenerative orphan diseases

PRODUCT PIPELINE

	Designations				
Neuropathic lysosomal diseases	Orphan Drug	Fast Track	Breakthrough Therapy		**Key milestone**
Niemann-Pick disease Type C*	✓	✓	✓	Ph 2/3 (data reported)	Regulatory submissions (US H1 2020 / EU H2 2020)
Neurological Gaucher disease				Ph 2	Ph 2 results H1 2020
GCase-Parkinson's disease**				Pre-clinical	
Neuromuscular disorders					
Sporadic Inclusion Body Myositis	✓	✓		Ph 2/3	Ph 2/3 results H1 2021
Amyotrophic Lateral Sclerosis	✓			Ph 3	Ph 3 results H1 2021

*Arimoclomol has been granted Rare Pediatric Disease Designation by the FDA for NPC, **Glucocerebrosidase (GCase)

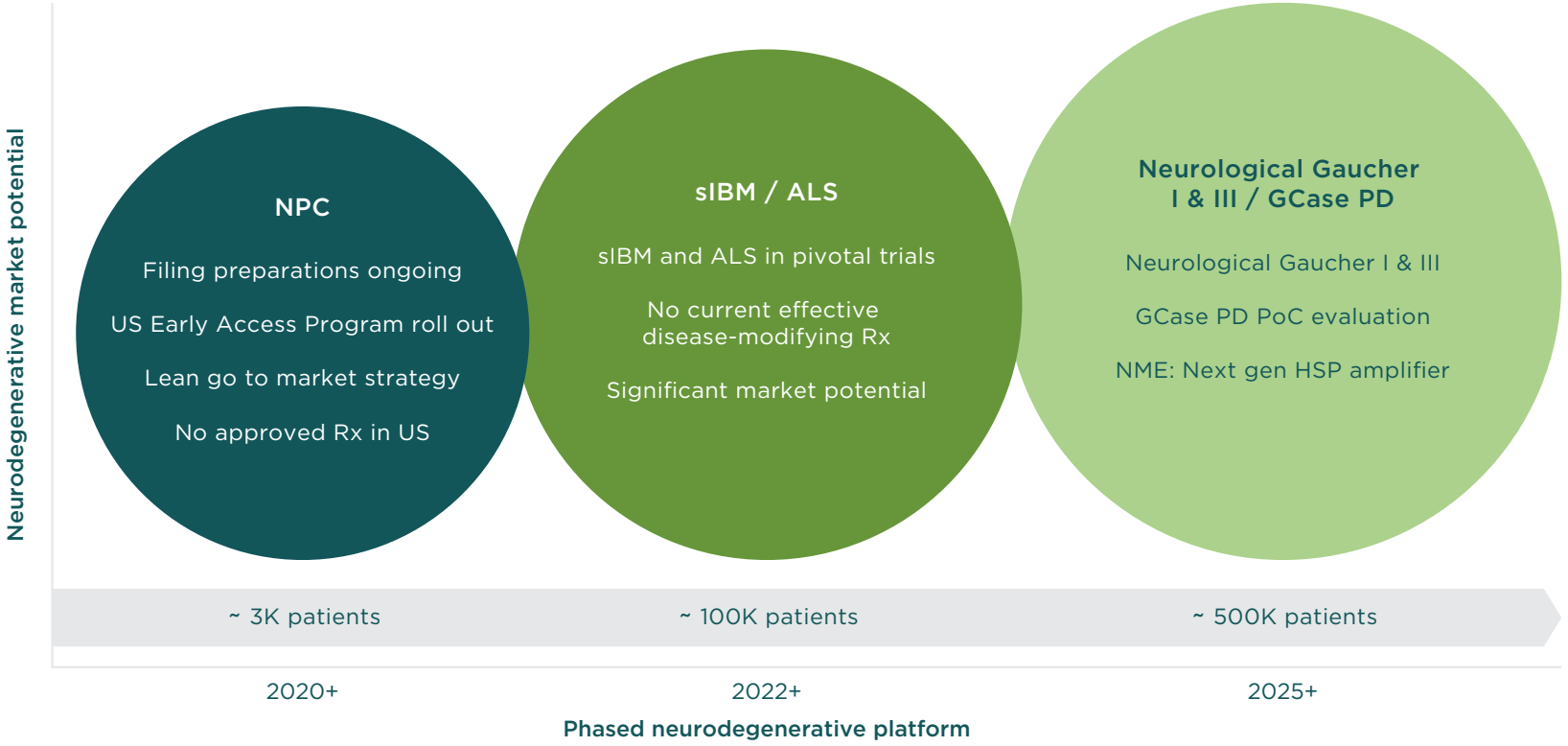
ARIMOCLOMOL PIPELINE

First-in-class HSP amplifier
Our lead investigational product candidate, arimoclomol, is a first-in-class Heat-Shock Protein (HSP) amplifier. HSPs are a key player in the Heat-Shock response, a cell's natural cellular stress defense system.

Significant potential in a range of neurodegenerative diseases
HSP amplification has been demonstrated to correct protein misfolding, break down protein aggregates, and improve lysosomal function, which are key requirements in a broad range of neurodegenerative diseases.

Ability to target CNS
Arimoclomol has the ability to cross the blood-brain barrier. This enables the product candidate to reach the source of neurological disease and creates additional opportunities for use in diseases such as ALS, sIBM, neurological Gaucher disease, and certain forms of Parkinson's disease (PD).

Attractive product profile
Arimoclomol is orally available, and can be administered by mouth, as a pill, or through a gastric tube. No significant safety concerns have been observed in ~540 patients and healthy volunteers treated to date.



NIEMANN-PICK DISEASE TYPE C

Niemann-Pick disease Type C (NPC) is a rare, inherited, progressive, and often fatal neurodegenerative disease. NPC is a lysosomal storage disorder caused by genetic mutations that most often lead to misfolded variants of the NPC proteins.

Misfolded NPC protein does not function properly and is subject to rapid degradation. As a consequence, lipids, that would normally be cleared, build up in the lysosomes of cells throughout the body. Accumulation of lipids in the tissues and organs, including the brain, leads to loss of cell function and organ damage. Neurologic involvement is common and results in progressive motor and cognitive impairment.

NPC is caused by mutations on one of two genes, NPC1 or NPC2. Approximately 95% of individuals with the disease have mutations in NPC1.

TREATMENT OPTIONS

The majority of current treatment options are palliative and are only directed towards the specific symptoms apparent in each individual (e.g. prescription of anti-seizure medications to prevent seizures). Only one drug, Zavesca® (miglustat), is currently marketed for NPC and only in certain jurisdictions. The product may reduce progression of disease in some patients, but there is still a very large unmet need for new therapy in NPC.

PREVALENCE

NPC often appears in childhood but can appear at any age. The incidence of the disease is estimated to be 1 in 100,000 live-births and it is estimated that there are ~2,000 patients across EU and US.

TRIAL STATUS

Positive data from a randomised, placebo-controlled phase 2/3 study evaluating arimoclomol on top of routine clinical care has been reported, along with additional data from a 12-month open-label extension study; arimoclomol continued to slow disease progression over two years. Patients who initiated treatment with arimoclomol in the open-label extension experienced a 90% reduction when comparing to the previous year on randomized placebo-treatment. We plan to submit arimoclomol for approval in the US in H1 2020 and in Europe in H2 2020 for NPC, with potential approvals in H1 2021 and H2 2021, respectively.

EARLY ACCESS PROGRAM

Orphazyme is deeply committed to providing access to new therapies for people living with rare diseases such as NPC. In January 2020, Orphazyme launched an Early Access Program (EAP) for arimoclomol in NPC in the US. Sometimes called "compassionate use," expanded, or early access provides a pathway for patients with serious, life-threatening diseases or conditions who lack therapeutic alternatives to gain access to investigational drugs before they are approved.

There are currently no approved products to treat NPC in the US and we are proud to make arimoclomol available pre-commercially to US patients through this program. We are currently evaluating how to offer early access in additional countries over time, contingent upon discussions with local authorities and our progress towards filing for regulatory approval or obtaining reimbursement.

Orphazyme has partnered with Clinigen Group to administer the arimoclomol EAP and support physicians interested in participation. The EAP is expected to remain open until arimoclomol becomes commercially available in the US.

80% reduction in disease progression

when individuals with double functional null mutations are excluded

AMYOTROPHIC LATERAL SCLEROSIS

Amyotrophic Lateral Sclerosis (ALS), also called Lou Gehrig's disease, is a rare neuromuscular disease, which is rapidly progressive and fatal, usually within two to five years. The disease attacks the neurons responsible for controlling muscles leading to paralysis of all skeletal muscles, eventually also affecting breathing, speaking, and swallowing.

The cause of damage to the neurons includes protein misfolding and aggregation.

FAMILIAL AND SPORADIC ALS

Approximately 10% of ALS cases are associated with familial ALS, while the rest have no identified genetic component (sporadic ALS). Amongst familial ALS cases, 20% harbor mutations in a SOD1 enzyme. SOD1 ALS is often very aggressive with a life expectancy of less than 2 years from diagnosis.

Arimoclomol has so far been tested in two phase 2 ALS trials, one dose-ranging trial in sporadic ALS, and one trial in ALS caused by SOD1 mutations.

TREATMENT OPTIONS

There are currently very limited treatment options available to ALS patients, namely; Rilutek® (riluzole) which reduces the levels of the neurotransmitter glutamate, potentially slowing down the progression of ALS; and Radicava® (edaravone) which can act against the reactive oxygen species that may damage the nerve cells in ALS patients. A high unmet medical need remains for new therapies for ALS patients.

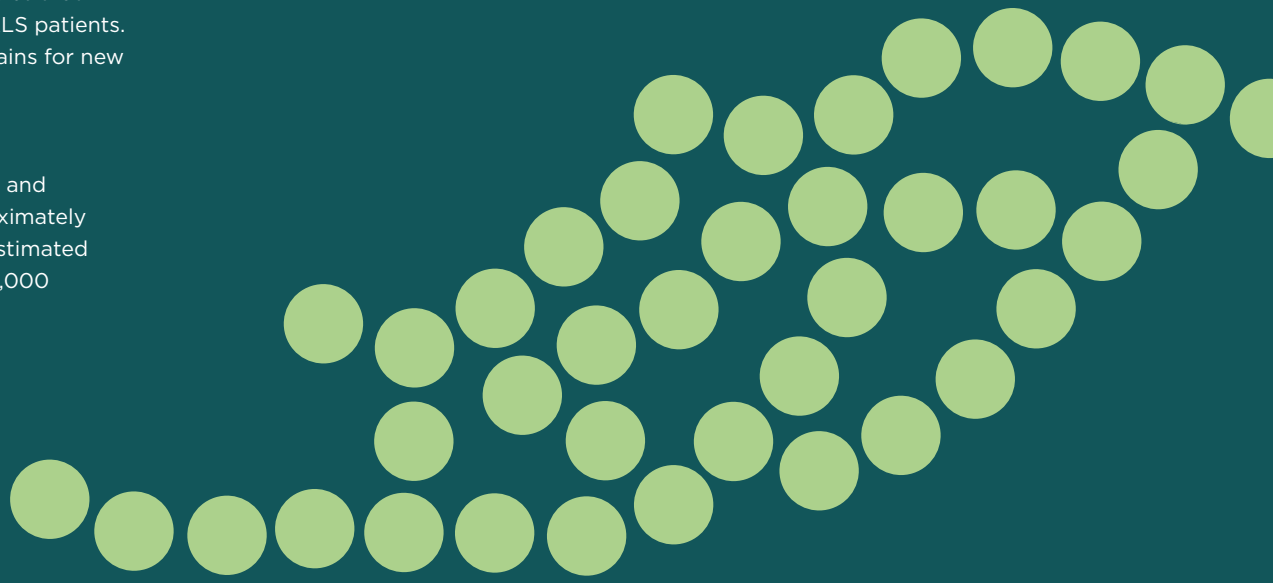
PREVALENCE

The patient population in the US and Europe is estimated to be approximately 50,000 patients. In Japan, it is estimated that there are between 8,000-14,000 patients with ALS.

TRIAL STATUS

In July 2019, Orphazyme completed the enrollment of 213 patients at 32 trial sites for a phase 3 trial in the US and Europe to support the application for a marketing authorization in ALS. Top-line results are expected in H1 2021.

Up to 50,000 patients in US and Europe



SPORADIC INCLUSION BODY MYOSITIS

Sporadic Inclusion Body Myositis, or sIBM, is an acquired, rare, and slowly-progressing protein-aggregation disease. It is the most common muscle-wasting disorder in the elderly population and is characterized by progressive degeneration, weakness, and atrophy of muscles, especially of the arms and legs.

In most cases, the disease progresses relentlessly over 10-15 years until the affected patient has lost mobility entirely. The cause of sIBM is not fully known, but degenerative factors, i.e. the build-up of tangled and misfolded proteins (inclusion bodies), play a major role.

TREATMENT OPTIONS

There is currently no approved drug for the treatment of sIBM. The standard treatment option for sIBM consists only of supportive therapy (physical, speech, and occupational therapy), and there is no evidence that any form of treatment slows progression.

PREVALENCE

sIBM is the most common idiopathic inflammatory myopathy in people over 50 years. The size of the sIBM patient population in the US and Europe is not fully known, but it is estimated that there are up to 40,000 patients suffering from this disease.

TRIAL STATUS

A phase 2/3 trial evaluating arimoclomol in sIBM is being conducted at 12 sites in the US and Europe and is now fully enrolled. The trial is intended to support registration of arimoclomol for the treatment of sIBM. Top-line results are expected in H1 2021.

Up to 40,000 patients in US and Europe

Time to walking stick ~5 years

Time to wheel chair ~10-15 years

GAUCHER DISEASE

Gaucher disease is an inherited metabolic disorder caused by mutations in a protein called glucocerebrosidase, which leads to the accumulation of certain sugar-containing lipids. The usual symptoms of Gaucher disease include an abnormally enlarged liver and/or spleen (hepatosplenomegaly), low levels of circulating red blood cells (anemia), blood cells promoting clotting (thrombocytopenia), and skeletal abnormalities. Disease of the nervous system is observed in a significant subpopulation of Gaucher disease (neuronopathic Gaucher).

There are three main types of Gaucher disease: Type I is the most common form of the disease in Western countries; type II (acute infantile neronopathic Gaucher disease) and type III (chronic neuronopathic Gaucher disease) involve a significant neurological element which for type II patients is often fatal in the first few years of life.

TREATMENT OPTIONS

Two types of treatment are currently available for patients with Gaucher disease: Enzyme-replacement therapy and substrate reduction therapy. None of these are useful in the treatment of neuropathic Gaucher disease.

PREVALENCE

It is estimated that there are up to 15,000 Gaucher disease patients in the US and Europe. Up to 50% of patients with type I Gaucher disease develop neurological symptoms and 100% of patients with type III Gaucher have CNS (central nervous system) involvement. We are focused on these patients with neuronopathic Gaucher disease, for which no treatments are available today.

Up to 15,000 patients in US and Europe

TRIAL STATUS

A phase 2 clinical trial of arimoclomol in Gaucher disease was initiated in Q2 2018 and is fully enrolled. Results are expected in H1 2020.

No treatments are currently available for neuronopathic Gaucher disease

PARTNERSHIPS

UNIVERSITY OF KANSAS AND UNIVERSITY COLLEGE LONDON

License Agreement

In October 2017, Orphazyme entered into a license agreement with University of Kansas and UCL Business PLC (a wholly-owned subsidiary of University College London). The license agreement grants Orphazyme the world-wide, royalty-bearing exclusive license to develop and commercialize products under all data generated in the course of the on-going phase 2/3 clinical trial of arimoclomol for the treatment of sIBM. Orphazyme's license includes any inventions and know-how included in such data. The trial was initiated in August 2017 with the University of Kansas as sponsor, but the license agreement provides that the Investigational New Drug (IND) and trial sponsorship shall be transferred to Orphazyme on Orphazyme's request. Under the terms of the license agreement, Orphazyme shall pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme is required to use commercially diligent efforts to develop and commercialize such products. The license agreement also provides that Orphazyme in consideration of the license shall issue bonus shares in favor of the University of Kansas and UCL Business PLC, for up to an aggregated value of USD 2.5 million (around DKK 15.8 million) in total depending on the size of the grants awarded to the universities under the trial (with a price per share calculated based on the average closing price of the shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance). The shares shall be issued or delivered on a yearly basis subject to certain reporting requirements.

UNIVERSITY OF MIAMI

Option and License Agreement

In May 2017, Orphazyme entered into an agreement with the University of Miami granting it first option to negotiate an exclusive-worldwide license regarding arimoclomol in ALS (refer to Orphazyme's Prospectus, November 2017 for details). Orphazyme exercised its option in September 2019 and entered into a global royalty-bearing, exclusive license to use or apply data, know-how, and patent rights generated by the University of Miami in a phase 2 clinical trial of arimoclomol in ALS with the SOD1 mutation. Orphazyme paid USD 75K (TDKK 508) cash up-front and agreed to future payments of certain license fees, a milestone upon regulatory approval in ALS, and annual fees as well as a 0.75% royalty on net sales of products sold within ALS linked to SOD1 mutations. Annual fees will be creditable against royalty and milestone payments.

CYTRX

Asset Purchase Agreement

In May 2011, Orphazyme entered into an Asset Purchase Agreement with the US biopharmaceutical company CytRx. Pursuant to this agreement, CytRx irrevocably sold and transferred certain preclinical and clinical data, intellectual property rights, and other assets, including contractual rights and obligations relating to a portfolio of chemical compounds, including arimoclomol, to Orphazyme. Under the terms of the Agreement, Orphazyme paid USD 150K cash up-front and agreed to make future payments to CytRx contingent upon achievement of specified milestones and royalties on a specified percentage of net sales of products containing one of the compounds purchased. For NPC, Orphazyme shall, upon approval of arimoclomol by the FDA and/or EMA, pay, respectively, USD 6 million and USD 4 million to CytRx.

FINANCIAL REVIEW

INCOME STATEMENT

The net result for the financial year 2019 was a net loss of DKK 337.5 million compared to DKK 229.6 million in 2018. The increase is primarily due to a continued increase in research and development activities, the hiring of new employees, as well as production of arimoclomol.

RESEARCH AND DEVELOPMENT COSTS

Research and development expenses totaled DKK 285.4 million in 2019 compared to DKK 196.5 million in 2018. The increase was mainly due to the ramp-up of the sIBM phase 2/3 trial and the phase 3 trial for ALS, as well as the initiation of open-label extensions for these trials. In addition, we initiated pre-clinical trials in preparation for filing the NDA/MAA in 2020. The increase in clinical trial activities also demanded increased amounts of the drug substance arimoclomol. During 2019 we also grew our organization from 47 to 70 full-time R&D employees.

GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses totaled DKK 50.5 million in 2019 compared to DKK 35.1 million in 2018. The increase is mainly due to increased employee costs as our G&A employees increased from 10 to 16; the ramp-up of pre-commercial activities as well as increased Investor Relations activities. Employee costs are impacted by the hiring of new members of management and additional staff to some functions in-house.

NET FINANCIAL ITEMS

Net financials totaled an expense of DKK 7.0 million in 2019 compared to an income of DKK 3.4 million in 2018. The increase in financial expense is due to interest expense from the lease obligations recognized and the borrowings in 2019, which were not in place in 2018.

INCOME TAX BENEFIT

Income tax benefit totaled DKK 5.5 million in 2019 compared to DKK 5.5 million in 2018. The income tax benefit for the two years represents a tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act.

STATEMENT OF FINANCIAL POSITION

Cash

As of December 31, 2019, Orphazyme had cash DKK 123.6 million compared to DKK 394.7 million as of December 31, 2018. The decrease in cash results from the increase in spend as described above.

Equity

As of December 31, 2019, equity amounted to DKK 53.0 million compared with DKK 388.2 million as of December 31, 2018. The decrease mainly reflects the financial result for the year.

CASH FLOWS

Cash flow from operating activities

Net cash flow used in operating activities amounted to an outflow of DKK 326.8 million in the year ended December 31, 2019 compared to DKK 234.8 million in the year ended December 31, 2018. Net cash flow from operating activities is attributable primarily to the progression of clinical development activities, as well as general and administrative expenses.

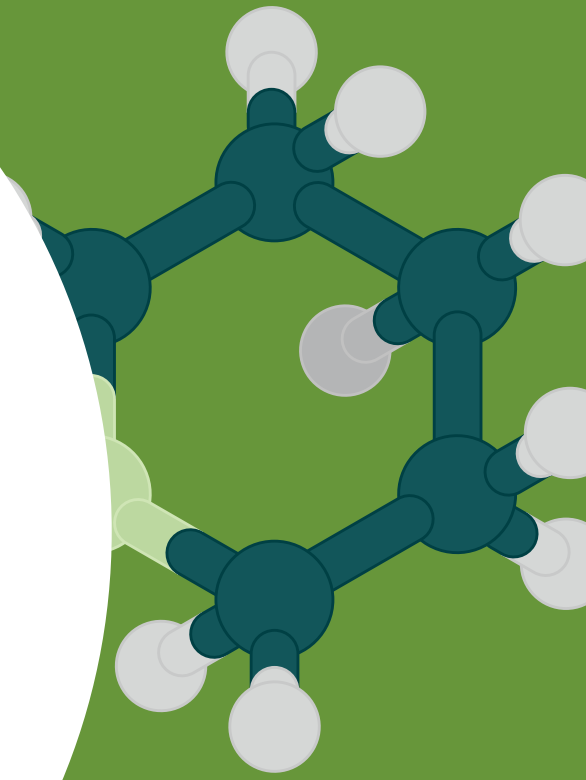
Cash flow from investing activities

Net cash outflow used in investing activities amounted to an outflow of DKK 3.3 million in the year ended December 31, 2019 compared to DKK 2.3 million in the year ended December 31, 2018. Investing activities comprise the purchase of equipment and the license agreement with the University of Miami.

Cash flow from financing activities

Net cash inflow from financing activities amounted to DKK 58.9 for the year ended December 31, 2019 compared to DKK 0 in the year ended December 31, 2018. The activity in 2019 is mainly due to the repayment of lease liabilities following adoption of IFRS 16 as well as the execution of the loan agreement resulting in a new inflow of cash.

Arimoclomol is a first-in-class Heat-Shock Protein amplifier



RISK MANAGEMENT

RISKS THAT THREATEN THE ACHIEVEMENT OF OUR KEY OBJECTIVES

Key objective
To successfully commercialize our product candidate in key markets.

Risks that threaten the achievement of our key objectives
Commercial risks include, but are not limited to: Our ability to obtain and maintain orphan designation/status, which will provide us with marketing exclusivity; our ability to establish in-house commercialization capabilities, including sales and marketing expertise in core markets; our ability to partner with third parties for distribution and/or commercialization; competition from other life science companies developing treatments for similar diseases, which could render our products obsolete or limit our ability to generate revenues; our ability to gain sufficient payor coverage and reimbursement; issues with manufacturing, availability, and supply of the product.

Our actions to mitigate the risks
In anticipation of positive feedback from discussions with regulatory authorities and eventual authorizations, we have started to ramp up our precommercialization and launch activities and potential commercial risks are assessed on an ongoing basis. Our commercial structure and operations are currently focused on NPC and on building out an expandable organization comprised of highly qualified talent with experience in the oversight and execution of product launches and commercial enterprises. Our new CEO, Kim Stratton, was hired to lead the commercialization efforts and comes with deep and relevant experience in this area.

Key objective
To successfully conduct and complete the ongoing clinical trials of arimoclomol in sIBM, ALS, NPC, and Gaucher disease and gain regulatory approvals required for commercialization of our products.

Risks that threaten the achievement of our key objectives
Designing and conducting clinical trials is complex, costly, and time-consuming and neither the results nor timing can be predicted with any certainty. There is a risk that clinical trial results will not confirm previous results, will produce adverse or inconclusive results or they will not demonstrate sufficient evidence of safety and efficacy to ensure the requisite regulatory approvals for marketing and sale. There is a risk that additional clinical trials or data may be required to obtain such approvals, which would result in increased costs, significant delays to filing with regulatory authorities, filing for a narrower indication than previously anticipated or the abandonment of efforts to commercialize one or more of the Company's product candidates.

Our actions to mitigate the risks
We make every effort to design and plan our clinical trials in the most diligent manner and perform them under strict regulatory guidelines. We partner with professional organizations to conduct our clinical trials and we conduct inspections and internal quality audits to maximize quality, safety and efficacy. We maintain frequent interactions with regulatory authorities to ensure that we are moving in the right direction to advance our programs towards approval in the most expedient manner.

Key objective
To use our expertise, including proprietary know-how, to select and develop new molecular entities (NMEs) for other rare, neurodegenerative diseases.

Risks that threaten the achievement of our key objectives
To a large extent, our success depends on our ability to obtain and maintain patents and other intellectual property rights for our products. Our IP is the basis for our current products and any potential new leads, and thus any threats to our IP rights could be detrimental to our future pipeline of product candidates.

Our actions to mitigate the risks
We are developing a suite of new molecular entities (NMEs) and have attracted highly talented resources to continue to develop and explore new leads. In addition, we are consistently monitoring our IP in order to not only protect our rights and minimize legal claims, but also strengthen our rights and current technology platform. We believe that our patent portfolio has a wide scope of protection and geographical coverage.

In addition to risks threatening the achievement of our key objectives, we are exposed to pervasive risks that threaten our business.

PERVASIVE RISKS THAT THREATEN OUR BUSINESS

Pervasive risk
Lack of sufficient financing.

How the risk threatens our business
We have not received approval for any product candidate for commercial sale and, as a result, have incurred significant financial losses, and may continue to incur significant financial losses in the future, which makes it difficult to assess the future viability of the Company. Furthermore, in order to execute our strategy, we may need to raise additional capital in the future and such funding may not be available on favorable terms. Failure to generate or secure sufficient financial resources could have a material adverse effect on the Company's business and/or prospects.

Our actions to mitigate the risks
In February 2020, we raised additional capital through a directed share issue and private placement. This is expected to cover our financing needs until well into 2021 and support filings for approval of arimoclomol for NPC, as well as preparations for commercial launch. Financial situation and risks are assessed on an ongoing basis and reported to the Audit Committee and the Board of Directors.

Pervasive risk
Non-compliance with legislation and industry standards.

How the risk threatens our business
We are subject to regulatory and legislative obligations in order to conduct business. These requirements are subject to change and if we do not remain abreast of the regulations and actively work to comply, we are at risk of either losing or not obtaining required approvals or we may receive penalties, fines, or suspension of our approvals or registrations. There is also a risk that cybersecurity attacks could compromise data privacy or cause interruption to our operations.

Our actions to mitigate the risks
Our organization has increased regulatory resources to facilitate interactions with regulators, actively monitor the current regulatory environment and ensure our compliance. We have implemented new IT-security procedures in order to reduce the risk of cybercrime. We have internal training requirements for all employees and contracting external suppliers, we ensure they have adequate measures in place to comply with relevant regulatory requirements.

Pervasive risk
Attraction and retention of talent.

How the risk threatens our business
The success of our company depends on our ability to attract, integrate, manage, and retain qualified personnel or key employees. Failure to do so could have a material adverse effect on the Company's business, results of operations, cash flows, financial condition, and/or prospects. The market for qualified personnel is competitive and the Company may not succeed in recruiting personnel to, for instance, commercialize its products as currently envisaged, or it may fail to effectively replace current personnel who depart with qualified or effective successors.

Our actions to mitigate the risks
We believe we have established an attractive workplace at Orphazyme, underpinned by high ethical standards. We are committed to maintaining a working environment that is diverse, free of discrimination, harassment, and bullying. We have established initiatives that provide opportunities for personal and professional development, improved health-wellbeing, work-life flexibility, and a participation in the overall success of Orphazyme through our long-term incentive plans.

CORPORATE GOVERNANCE

In order to maintain the trust of the Company's stakeholders, Orphazyme is committed to ensuring transparent and good corporate governance. As a company listed on Nasdaq Copenhagen, Orphazyme is subject to the Recommendations on Corporate Governance from November 2017.

The Recommendations on Corporate Governance are best practice guidelines for the management of companies admitted to trading on a regulated market.

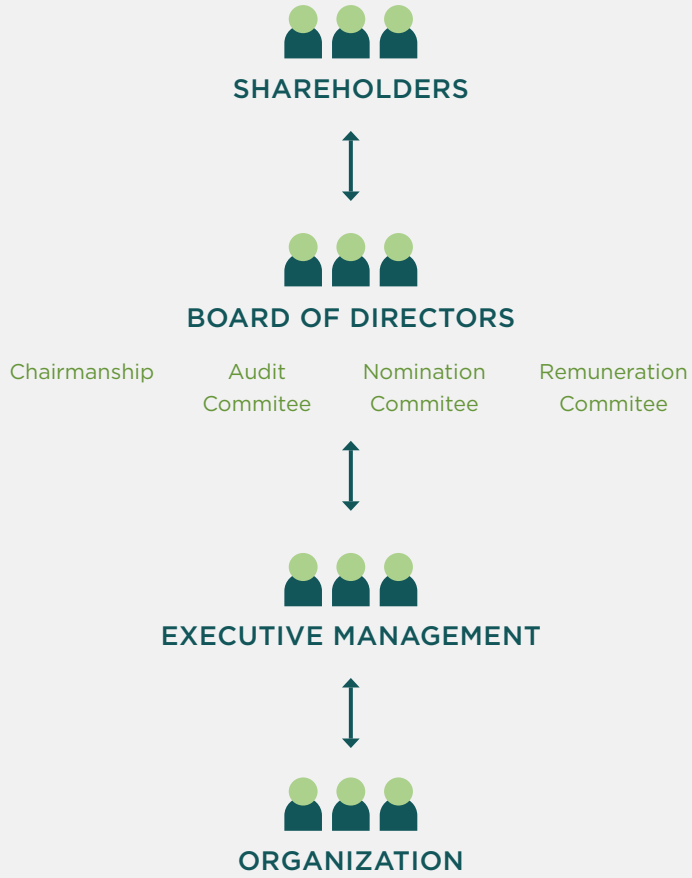
Orphazyme intends to comply with the Recommendations on Corporate Governance in all material respects, however, due to the current size of the Company and the nature of its present operations, the company has opted to deviate from the recommendations in the following areas:

- Orphazyme has decided to only publish annual reports and half-yearly financial reports;
- the annual report does not include information on individual board members' participation in board meetings;
- the general conclusion of the latest evaluation of the Board of Directors is not described in the annual report, but is accounted for by the Chairman at the annual general meeting;
- share-based remuneration is offered to the Board of Directors;
- share-based instruments be granted to board members shall have a maturity of one year from the date of allocation;
- the Chief Executive Officer may under certain circumstances be entitled to receive remuneration related to the notice period, including severance pay, which exceeds two years' total remuneration; and
- Orphazyme has decided not to prepare a remuneration report until the company is required to do so pursuant to section 139(b) of the Danish Companies Act.

Orphazyme's corporate governance statement includes a summary of the Company's governance structure, a description of internal control and financial reporting procedures, Orphazyme's position on the Recommendation on Corporate Governance as well as a complete list of the Company's comments to recommendations that the Company opted to deviate from.

The corporate governance statement is available on www.orphazyme.com.

Corporate Governance Statement



BOARD OF DIRECTORS

Name	Position	Independent[1]	Year of first appointment	Expiration of term
Georges Gemayel	Chairman	Independent	2012	2020
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2020
Martin Bonde	Member	Independent	2010	2020
Sten Verland	Member	Independent	2010	2020
Rémi Droller	Member	Independent	2015	2020
Martijn Kleijwegt	Member	Independent	2017	2020
Catherine Moukheibir	Member	Independent	2017	2020
Anders Hedegaard	Member	Independent	2017	2020

[1] The Company has based its assessment of independence on the basis of criteria set out in the current Coporate Governance Recommendations (as defined below).

BOARD OF DIRECTORS

The Board of Directors is responsible for the overall and strategic management and proper organization of Orphazyme's business and operations and it supervises the Company's activities, management, and organization. The Board of Directors appoints and dismisses the members of the Executive Management, who are responsible for the day-to-day management of the Company.

The Board of Directors normally holds at least five regular meetings annually, including a strategy review, plus ad-hoc meetings as required. Extraordinary board meetings are convened by the Chairman when necessary or when requested by a member of the Board of Directors, a member of the Executive Management, or by the Company's auditor. The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman. Resolutions of the Board of Directors are passed by a simple majority of the votes present at the meeting. In the event of equal votes, the Chairman or, in his absence, the Deputy Chairman shall have the casting vote. The Board of Directors conducts an annual evaluation of the effectiveness, performance, achievements, and competencies of the Board of Directors and of the individual members as well as the collaboration with the Executive Management.

The members of the Board of Directors elected by the general meeting are elected for a term of one year. Members of the Board of Directors may be re-elected.

The Company believes that the members of the Board of Directors possess the professional skills and experience required to serve as board members of the Company.

BOARD COMMITTEES

To support the Board of Directors in its duties, the Board of Directors has established and appointed an Audit Committee, a Nomination Committee, and a Remuneration Committee. These committees are charged with reviewing issues pertaining to their respective fields that are due to be considered at board meetings. Written charters specifying the tasks and responsibilities for each of the committees are available on the Company's website www.orphazyme.com.

Committee Charters Summary

AUDIT COMMITTEE

The Audit Committee reviews accounting and audit matters that by decision of the Board of Directors or the Audit Committee require a more thorough evaluation, and assess the internal controls and risk management systems of Orphazyme. Its duties also include supervision of the Company's auditors and review of the audit process. In accordance with the Recommendations on Corporate Governance of the Danish Committee on Corporate Governance issued in November 2017 (the "Corporate Governance Recommendations"), the Company has decided that the Chairman of the Board of Directors may not also be the Chairman of the Audit Committee and that a majority of the members of the Audit Committee are required to meet the independence requirements set out in the Corporate Governance

© Copyright 2019 NPUK, this picture is kindly lent to Orphazyme A/S by NPUK

Recommendations. In addition, at least one member shall have accounting or audit qualifications and between them, the members shall possess such expertise and experience as to provide an updated insight into, and experience in, the financial, accounting, and audit aspects of companies with shares admitted to trading and official listing on a regulated market.

The Audit Committee shall consist of no less than three members appointed by and among the Board of Directors, including the Chairman of the Audit Committee, and consists of Catherine Moukheibir as Chairman, Martijn Kleijwegt, and Sten Verland. All of the members of the Audit Committee meet the independence requirement set out in the Corporate Governance Recommendations. The Chief Executive Officer and/or the Chief Financial Officer and the Company's external auditor shall participate in meetings of the Audit Committee if so requested by the Audit Committee and the external auditor shall attend at least one meeting per year or the relevant part hereof where the Executive Management is not present.

NOMINATION COMMITTEE

The Nomination Committee shall assist the Board of Directors with ensuring that appropriate plans and processes are in place for the nomination of candidates to the Board of Directors, the Executive Management, and the board committees. Moreover, the Nomination Committee shall evaluate the composition of the Board of Directors and the Executive Management. This includes making recommendations for nomination or appointment of members of (a) the Board of Directors, (b) the Executive Management, and (c) the board committees established by the Board of Directors.

The Nomination Committee consists of no less than three members appointed by and among the Board of Directors, including Georges Gemayel as Chairman, Martin Bonde, and Sten Verland. All of the members of the Nomination Committee meet the independence requirements set out in the Corporate Governance Recommendations.

REMUNERATION COMMITTEE

The Remuneration Committee ensures that the Company maintains a Remuneration Policy for the members of the Board of Directors and the Executive Management, and to evaluate and make recommendations for the remuneration of the members of the Board of Directors and the Executive Management.

The Remuneration Committee shall consist of no less than three members appointed by and among the Board of Directors, including Bo Jesper Hansen as Chairman, Rémi Droller, and Anders Hedegaard. All of the members of the Remuneration Committee meet the independence requirements set out in the Corporate Governance Recommendations.

DESCRIPTION OF INTERNAL CONTROL AND FINANCIAL REPORTING PROCEDURES

The Board of Directors, the Audit Committee, and the Executive Management are ultimately responsible for Orphazyme's risk management and internal controls in relation to its financial reporting and approve Orphazyme's general policies in that regard. The Audit Committee assists the Board of Directors in overseeing the reporting process and the most important risks involved in this respect. The Executive Management is responsible for the effectiveness of the internal controls and risk management and for the implementation of such controls aimed at mitigating the risk associated with the financial reporting. Orphazyme has internal control and financial reporting procedures aimed at enabling it to monitor its performance, operations, funding, and risk.

CORPORATE SOCIAL RESPONSIBILITY

As a biopharmaceutical company, we are aware of our responsibilities towards society, patients, stakeholders, and employees and we believe that our corporate behavior should always be of the highest ethical standard. When conducting our business, we strive to demonstrate respect for key moral principles and comply with international regulations and good practice guidelines to make sure that our product is safe and meets strict quality guidelines. As most of our business activities are outsourced, we work closely with our partners to ensure compliance with these regulations and guidelines.

We believe that our ethical behavior strengthens Orphazyme's status as an attractive workplace for both current and future employees.

Diversity Policy

BUSINESS MODEL

Orphazyme is a Danish biopharmaceutical company with approximately 90 employees. The office in Copenhagen serves as headquarters and focuses on coordination and execution of the drug development process and on administration of pre-clinical and clinical trials.

We are pioneering the Heat-Shock Protein (HSP) response for rare, neurodegenerative diseases. The key activity of Orphazyme is to develop innovative therapies for orphan diseases characterized by protein misfolding where we can apply our specialized know-how in HSPs. Our objective is to successfully conduct and complete the planned and ongoing trials of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC), sporadic Inclusion Body Myositis (sIBM), Amyotrophic Lateral Sclerosis (ALS), and Gaucher disease. We use external suppliers in order to both manufacture and tabletize the medicine, as well as for distribution.

RISK ANALYSIS

Risk is defined as the potential negative effect that can be experienced by the business or any of Orphazyme's stakeholders. Risk is seen as a combination between impact and likelihood of any given subject. Orphazyme's potential risk of impacting social and employee conditions, human rights, and anti-corruption is estimated to be limited, mainly due to the strictly regulated business environment the company operates in. Orphazyme's potential risk of impacting the environment and climate is estimated to be very limited mainly as a result of the modest number of employees and the use of external suppliers for all production. Orphazyme has prepared a policy for social and employee conditions, human rights, and anti-corruption. Insofar as a specific risk has been identified in any of these areas, it will be addressed by the relevant policy.

CSR REPORTING AREAS

ENVIRONMENT AND CLIMATE

Orphazyme does not have Environment or Climate policies. The Company acknowledges the challenges linked to climate change, and as a result Orphazyme operates an office focused on efficient management of office materials. However, based on the fact that the Company is operating from rented offices and conducts its business in a highly regulated industry and climate, Orphazyme's potential impact on the environment and climate is viewed as minimal. As a result, specific Environment and Climate policies have not been developed at this time.

HUMAN RIGHTS

Policies

Orphazyme acknowledges and supports the maintenance of internationally declared human rights and bases its work on the UN Universal Declaration of Human Rights and the interpretation that it is the responsibility of the State to protect, and the companies' responsibility to respect these rights.

Orphazyme conducts its business in accordance with the ethical and scientific principles governing clinical research on human subjects, as set out in the Declaration of Helsinki and the International Conference on Harmonization (ICH) guidelines on Good Clinical Practice (GCP).

Orphazyme interprets human rights to comprise respect for diversity. Therefore, it is the aim of the Company to ensure that employees have the possibility to function in a workplace without harassment regardless of gender, age, race, religion, or ethnicity.

Activities and results

All protocols in 2019 have been carried out in accordance with the current ICH-GCP guidelines. Furthermore, since 2018, Orphazyme has included in its supplier contracts a new condition that requires all suppliers to comply with the Declaration of Helsinki.

Orphazyme has a whistleblower policy in order to allow reporting of potential violations of the respect for diversity in the workplace. The policy was established in 2018 and mandatory meetings are held annually for all employees. In 2019, there have been no reports via the whistbleblower scheme concerning violations of the respect for diversity in the workplace or other human rights violations.

ANTI-CORRUPTION AND BRIBERY

Policies

Orphazyme is committed to maintaining the highest standards of ethical conduct and will not tolerate the use of bribery or corruption to achieve its business objectives. Employees must decline any expensive gifts, money, trips, or other such offerings from business contacts. This also includes receiving services from suppliers without paying for them.

Activities and results

Orphazyme has a whistleblower policy in order to allow reporting of potential violations of laws and serious violations of internal policies and procedures, including fraud and anti-corruption. The policy was established in 2018 and mandatory joint meetings are held annually for all employees. Orphazyme continued, via its Employee Handbook, to familiarize all new employees with the company policies regarding anti-corruption.

Orphazyme only enters into agreements with consultants that have signed the acknowledgement of Orphazyme's commitment to maintaining the highest standards of ethical conduct.

Orphazyme has not identified any cases of anti-corruption or bribery incidents throughout the year nor have any incidents been reported via the whistleblower scheme.

SOCIAL AND STAFF MATTERS

The office in Copenhagen serves as our headquarters and focuses on coordination and execution of the drug development process and on the conduct of preclinical and clinical trials administration. As of December 31, 2019, the organization comprised:

- Senior Management (5 persons comprising Executive Management and Key Employees)
- Administration (6 persons)
- Clinical development (42 persons)
- Chemistry, manufacturing, and controls/ quality assurance (2 persons)
- Regulatory (6 persons)
- Research (19 persons)
- Finance, legal, and IT (6 persons)

Policies

Orphazyme A/S knows that all employees are critical to the success of the Company and its programs. The Company is a diverse workplace committed to maintaining a working environment that is free of discrimination, harassment, and bullying. Orphazyme views diversity as an integrated part of a socially responsible company. In accordance with applicable law, we have adopted a Diversity Policy, which sets out our goals for increasing the diversity in the Board of Directors and at other management levels. We encourage diversity, including age, ethnicity, nationality, religion, education, and skills. Currently, our staff consists of 65.5% females and 34.6% males, including 67% female and 33% male employees at director level or above. The Board of Directors currently comprises one woman and seven men. The Board of Directors' target is to include at least two female board members by the end of 2021, which we intend to achieve. However, during 2019 we believe the board composition was ideally suited to our current activities and have not made changes.

Orphazyme values the personal development of its employees and offers employees the opportunity to participate in conferences and courses in order to strengthen their professional competences. Policies regarding staff matters are further elaborated in the Employee Handbook.

Activities and results

In 2019, Orphazyme continued to actively encourage all potential employees to apply for employment opportunities, irrespective of gender, age, race, religion, or ethnicity. Moreover, Orphazyme continues to introduce all new employees to the Employee Handbook in order to ensure a workplace that respects diversity and other values inherent in the Company.

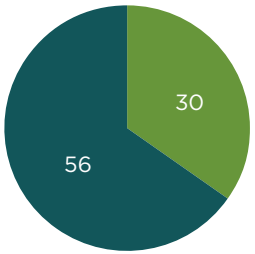
In 2019, Orphazyme continued its efforts to offer its employees opportunities for personal learning and development by providing internal training as well as external continuing education courses. The Company maintains its focus on a healthy work-life balance by encouraging employees to take all holidays to which they are entitled, and by providing opportunities such as working from home if needed. Furthermore, in 2019 Orphazyme continued to promote a healthy lifestyle by offering weekly employer-paid exercise options.

In 2019, Orphazyme has been able to retain a highly satisfactory employee turnover rate.

KEY EMPLOYEE RATIOS

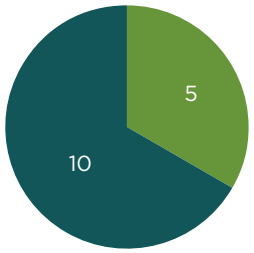
	2019		2018	
	Male	**Female**	**Male**	**Female**
Orphazyme A/S	35%	65%	35%	65%
Executive Management and Key Employees	80%	20%	100%	0%
Director level and above	33%	67%	30%	70%
Below director level	31%	69%	29%	71%

Year-end FTEs - 2019

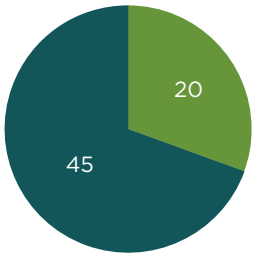


Director level and above - 2019

(not including Executive Management and Key Employees)



Below director level - 2019



Female Male

SHAREHOLDER INFORMATION

OWNERSHIP

Since November 16, 2017, Orphazyme has been listed on Nasdaq Copenhagen under the ticker symbol ORPHA.CO. In compliance with the rules and regulations stipulated for companies listed on this exchange, we conduct our communication in accordance with the Danish Financial Supervisory Authority and Nasdaq Copenhagen.

35%

ownership by major shareholders

As of December 31, 2019, the number of registered shareholders totaled 5,731 shareholders holding a total of 18,950,699 shares, which represented 94.8% of the total share capital of 19,984,799. In January 2020, there was a capital increase of 20,650 shares related to the issue of bonus shares to Kansas Life Sciences Development Company, Inc. and UCL Business PLC, and in February 2020 there was a capital increase of 7,032,937 shares, resulting in a total share capital of 27,038,386 as of February 28, 2020, the date of this annual report.

All shares have the same rights, including in respect of eligibility to receive dividends and participate in share buybacks. Orphazyme has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

MAJOR SHAREHOLDERS[1,2,3]

Major shareholder	Company address	Share capital %
LSP V Coöperatieve U.A.	Johannes Vermeer, Plein 9, 1071 DV Amsterdam, Netherlands	10.03%[4]
Novo Holdings A/S	Tuborg Havnevej 19, 2900 Hellerup, Denmark	7.5%
Sunstone Life Science Ventures Fund II K/S	Store Strandstræde 18 A, st., 1255 København K	6.67%
Coöperatieve Aescap Venture I U.A.	Barbara Strozzilaan 101, 1083 HN Amsterdam, Netherlands	6.53%

[1] Based on the latest major shareholder notification received by Orphazyme prior to publication of this Annual Report.
[2] In accordance with company announcement 18-2020, February 12, 2020, Consonance Capman GP LLC indirectly holds 1,900,000 shares and voting rights in Orphazyme, corresponding to 7.03% of the total share capital and voting rights of Orphazyme.
[3] In accordance with company announcement 19-2020, February 13, 2020, Danske Bank A/S directly and indirectly held 4.85% of the total share capital and 7.24% of the total voting rights.
[4] Includes a direct shareholding of 1.03% and an indirect shareholding of 9.00% of the total share capital and voting rights held through Orpha Pooling B.V. (a JV between LSP V Cooperative U.A. and ALS Invest 2 B.V.)

INVESTOR RELATIONS

Orphazyme's Investor Relations' primary goal is to ensure a timely communication of anything interesting or relevant to our stakeholders. This we do by communicating both company announcements (containing potentially share-sensitive information) and investor news (pertaining to interesting news that is not share-sensitive).

We strive to keep our shareholders and investors informed at all times, by providing annual and half-year reports, hosting analyst and investor meetings, upholding an informative and transparent website, containing all relevant reports, announcements, policies, etc. Orphazyme is followed by four analysts: Carnegie, Danske Equities, Oddo, and Redeye.

If you have any questions for Investor Relations, please feel free to contact Orphazyme's Chief Financial Officer, Anders Vadsholt: afv@orphazyme.com

Please visit our website for further details: www.orphazyme.com

SHARE PERFORMANCE

COVERING ANALYSTS

Carnegie, Danske Equities, Oddo, and Redeye

FINANCIAL CALENDAR

Annual General Meeting: Thursday, March 26, 2020
Interim Report H1 2020: Friday, August 28, 2020

NEWS

2019 COMPANY ANNOUNCEMENTS

30 **JANUARY 30**
Orphazyme reports positive results from full data set of Phase 2/3 arimoclomol trial in Niemann-Pick disease Type C (NPC)

Major shareholder announcement

31 **JANUARY 31**
Capital increase of 26,060 shares (equivalent to approximately 0.13% of the existing shares) in Orphazyme A/S as a result of an issue of bonus shares to KLSDC and UCLB

1 **MARCH 1**
Orphazyme announces Annual Report 2018

Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

5 **MARCH 5**
Notice to convene Annual General Meeting

Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

7 **MARCH 7**
Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

8 **MARCH 8**
Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

27 **MARCH 27**
Resolution passed at the Annual General Meeting

29 **MARCH 29**
Share capital and voting rights in Orphazyme A/S

17 **APRIL 17**
Major shareholder announcement

23 **APRIL 23**
Orphazyme's Phase 2/3 trial in sporadic Inclusion Body Myositis fully enrolled

31 **MAY 31**
Major shareholder announcement

7 **JUNE 7**
Orphazyme to prepare for filing of arimoclomol in Europe for Niemann-Pick disease Type C (NPC)

NEWS

2019 COMPANY ANNOUNCEMENTS

15 **JULY 15**
Orphazyme appoints Kim Stratton as Chief Executive Officer, effective October 1, 2019

18 **JULY 18**
Orphazyme completes enrollment in phase 3 trial evaluating arimoclomol in Amyotrophic Lateral Sclerosis

21 **JULY 21**
Orphazyme to prepare for filing of arimoclomol in US for Niemann-Pick disease Type C (NPC)

26 **JULY 26**
Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

29 **JULY 29**
Reporting of transactions in Orphazyme's shares made by persons discharging managerial responsibilities

27 **AUGUST 27**
Orphazyme strengthens balance sheet with EUR 9 million financing from Kreos Capital

2 **OCTOBER 2**
Major shareholder announcement

1 **NOVEMBER 1**
Financial calendar 2020

19 **NOVEMBER 19**
Orphazyme receives Breakthrough Therapy Designation for arimoclomol in Niemann-Pick Disease Type C (NPC)

28 **NOVEMBER 28**
Major shareholder announcement

18 **DECEMBER 18**
Orphazyme's arimoclomol receives US Fast Track designation in sporadic Inclusion Body Myositis

19 **DECEMBER 19**
Major shareholder announcement

NEWS

2019 INVESTOR NEWS

25

FEBRUARY 25

Orphazyme Annual Report 2018 presentation

11

MARCH 11

Orphazyme to present at Cowen & Co Annual Health Care Conference

21

AUGUST 21

Orphazyme Interim Report First Half 2019 presentation

17

SEPTEMBER 17

Orphazyme CMO to speak at FDA-funded rare disease data analytics platform launch event

BOARD OF DIRECTORS

GEORGES GEMAYEL

Chairman of the Board

Member since: 2012 (Chairman, 2014)
Born in: 1960
Nationality: American
Committees: Nomination (Chairman)

Special competencies:
Dr Gemayel has significant management and executive experience from the global pharmaceutical industry. He holds a Master's and PhD in Pharmacology from Paris-Sud University and a Docteur d'Exercice en Pharmacie, St. Joseph University.

Current positions:
Chairman of the Board of Dynacure, Enterome SA, and OxThera AB, and a member of the Board of Directors of Momenta Pharmaceuticals Inc. (publ) and Supernus Pharmaceuticals Inc. (publ).

BO JESPER HANSEN

Deputy Chairman of the Board

Member since: 2010 (Deputy Chairman, 2017)
Born in: 1958
Nationality: Danish
Committees: Remuneration (Chairman)

Special competencies:
Dr Hansen has extensive experience in orphan drugs, both from the operations and supervisory point of view and has broad and current know-how of the biotechnology environment. He holds an MD and a PhD in Medicine from the University of Copenhagen.

Current positions:
Chairman of the Board of Laborie Inc., Innoventa Medica ApS, Karo Pharma AB; member of the Board of Azanta A/S and Ascelia Pharma AB. Venture Partner at Wellington Partners; Advisory Consultant for Aescap 2.0, Nordic Capital, EQT AB and Broad Street Principal Investments Europe Ltd. & senior business advisor for HBM Ventures Ltd.

MARTIN BONDE

Member since: 2010
Born in: 1963
Nationality: Danish
Committees: Nomination

Special competencies:
Dr Bonde has executive experience and in-depth knowledge of the biotechnology environment in the Nordic countries. He holds a Graduate Diploma in Business Administration from Copenhagen Business School, an MSc. and a PhD in Chemical Engineering from the Technical University of Denmark.

Current positions:
Entrepreneur-in-Residence at BiOrigin ApS, a Novo Seeds company; member of the Board of Directors of Visiopharm A/S, Chief Executive Officer of Bohrs Tower ApS as well as a member of the Board of Directors and the Executive Management of Biotopix ApS.

RÉMI DROLLER

Member since: 2015
Born in: 1975
Nationality: French
Committees: Remuneration

Special competencies:
Mr Droller has extensive experience as a biotechnology investor and a proven track-record in negotiating several successful transactions. He holds a Master's in Molecular Biology from Université Pierre et Marie Curie and a Master's in Finance and Management of Innovation from AgroParisTech.

Current positions:
Managing Partner of Kurma Partners SA and member of the Board of Directors of Dynacure SAS, ImCheck Therapeutics SAS, OxThera AB, AM Pharma BV, Flamingo Therapeutics BV, Vico Therapeutics BV, and Pharvaris BV.

BOARD OF DIRECTORS

MARTIJN KLEIJWEGT

Member since: 2017
Born in: 1955
Nationality: Dutch
Committees: Audit

Special competencies:
Mr Kleijwegt has extensive experience as a major European venture-capital investor as well as in-depth experience from the pharmaceutical industry. He holds a Master's degree in Economics from the University of Amsterdam.

Current positions:
Founder and Managing Partner at LSP Management Group BV and a member of the Board of Directors of AM Pharma BV, Kiadis Pharma N.V. (publ), OxThera AB, Eloxx Pharmaceuticals Ltd., and Pharvaris BV.

STEN VERLAND

Member since: 2010
Born in: 1957
Nationality: Danish
Committees: Nomination, Audit

Special competencies:
Dr Verland is a serial entrepreneur in biotechnology companies and has extensive investment and managerial experience. He holds a Master's in Biology and Mathematics and a PhD in Immunology from the University of Copenhagen.

Current positions:
Co-Founder and Board Member at Sunstone Capital A/S, Board Member and General Partner at Sunstone Life Science Ventures A/S, Board Member of STipe Therapeutics ApS, Anergis SA, MinervaX ApS, OxThera AB, the Danish VC and PE Association (DVCA), Board Member and Executive Management in certain Sunstone Group companies; member of the Executive Management at Verland Capital ApS, Verland Holding ApS, Verland Holding II ApS, and Genobiotix ApS.

CATHERINE MOUKHEIBIR

Member since: 2017
Born in: 1959
Nationality: American, Lebanese, British
Committees: Audit (Chairman)

Special competencies:
Ms Moukheibir has in-depth experience in the pharmaceutical and banking industries and a successful track-record in leading Audit Commitees of publicly-traded companies. She holds a Master's degree in Economics and an MBA, both from Yale.

Current positions:
Chairman of the Board of Directors and Chief Executive Officer of MedDay Pharmaceuticals. Member of the Board of Directors of Genkyotex SA (publ), Ironwood Pharmaceuticals, Inc. and Kymab Ltd; member of the Advisory Board of Imperial College Business School.

ANDERS HEDEGAARD

Member since: 2017
Born in: 1960
Nationality: Danish
Committees: Remuneration

Special competencies:
Mr Hedegaard has extensive knowledge of the healthcare industry, both in product development and commercialization. Master of Science in Chemical Engineerng and Biochemistry from the Technical University of Denmark.

Current positions:
Anders Hedegaard is currently Chief Executive Officer of Rodenstock Group.

MANAGEMENT

EXECUTIVE MANAGEMENT

KIM STRATTON
Chief Executive Officer

Kim joined Orphazyme as Chief Executive Officer in October 2019.

Kim has more than 25 years' global commercial experience from biopharmaceuticals. Previously at Shire Pharmaceuticals and Novartis.

Kim Stratton is currently a member of the Board of Directors of Novozymes A/S (publ) and Vifor Pharma AG.

Kim is a Registered Nurse and received her certification at Royal North Shore Hospital, Australia.

Born in: 1962
Nationality: Australian

ANDERS VADSHOLT
Chief Financial Officer

Anders joined Orphazyme in May 2016 as Chief Financial Officer.

Anders has 20+ years' experience from biotech and corporate finance. Previously at Topotarget, BankInvest Biomedical Venture, 7TM Pharma, and Carnegie.

Anders is currently a member of the Board of Directors at OxThera AB and Owner of Alpha Healthcare Investments ApS.

Anders holds a BSc in Corporate Law from the University of Aalborg, an MBA in Finance and Strategy from the University of Melbourne, and an MSc in Corporate Law and Economics from Copenhagen Business School.

Born in: 1969
Nationality: Danish

KEY EMPLOYEES

THOMAS KIRKEGAARD JENSEN, PHD
Chief Scientific Officer, Co-Founder

Thomas joined the Company as Co-Founder and Chief Executive Officer in 2009 and became Chief Scientific Officer in March 2010.

Thomas is currently a member of the Executive Management of Dare to Dream ApS, an expert reviewer for the European Research Council and a member of the Advisory Board for the Rare Disease Report.

Thomas holds a BSc in Biochemistry, an MSc in Human Biology and a PhD in Medicine from the University of Copenhagen.

Born in: 1977
Nationality: Danish

THOMAS BLAETTLER, MD
Chief Medical Officer

Thomas joined Orphazyme in November 2016 as Chief Medical Officer.

Thomas has 13+ years' experience in neuroscience development. Previously at Roche, Bristol-Myers Squibb, and Novartis.

Thomas Blaettler holds a Doctorate in Medicine from the University of Zürich and a Medical School Certificate Swiss State Examination from the Medical School of the University of Zürich. He is a board-certified neurologist by the Swiss Medical Association (the Foederation Medicorum Helveticorum).

Born in: 1967
Nationality: Swiss and Danish

GLOSSARY

Amyotrophic Lateral Sclerosis (ALS)
Also called Lou Gehrig's disease; a rare neuromuscular disease, which is rapidly progressive and fatal, usually within two to five years.

Breakthrough Therapy Designation (BTD)
FDA designation that is intended to expedite the development and review of drugs to treat serious and life-threatening diseases in cases where preliminary evidence shows that the drug may provide substantial improvements over available therapy.

Early Access Program (EAP)
Provides patients with serious, life-threatening diseases or conditions with unmet medical needs access to investigational drugs before they are approved.

European Medicines Agency (EMA)
Regulatory agency in Europe that facilitates development and access to medicines, evaluates applications for marketing authorization and monitors the safety of medicines.

Fast-Track Designation
FDA designation that is intended to facilitate the development and expedite review of drugs for serious diseases with an unmet medical need, getting new drugs to patients earlier.

Gaucher disease
An inherited metabolic disorder caused by mutations in a protein called glucocerebrosidase, which leads to the accumulation of certain sugar-containing lipids.

Heat-Shock Proteins
Heat-Shock Proteins (HSPs) are molecular chaperones constituting a natural system that makes other proteins work correctly and guard against toxicity arising from misfolded proteins and dysfunctional cellular recycling systems.

Sporadic Inclusion Body Myositis (sIBM)
An acquired, rare, and slowly-progressing protein-aggregation disease.

Marketing Authorization Application (MAA)
A submission to apply for marketing approval for a drug from EMA.

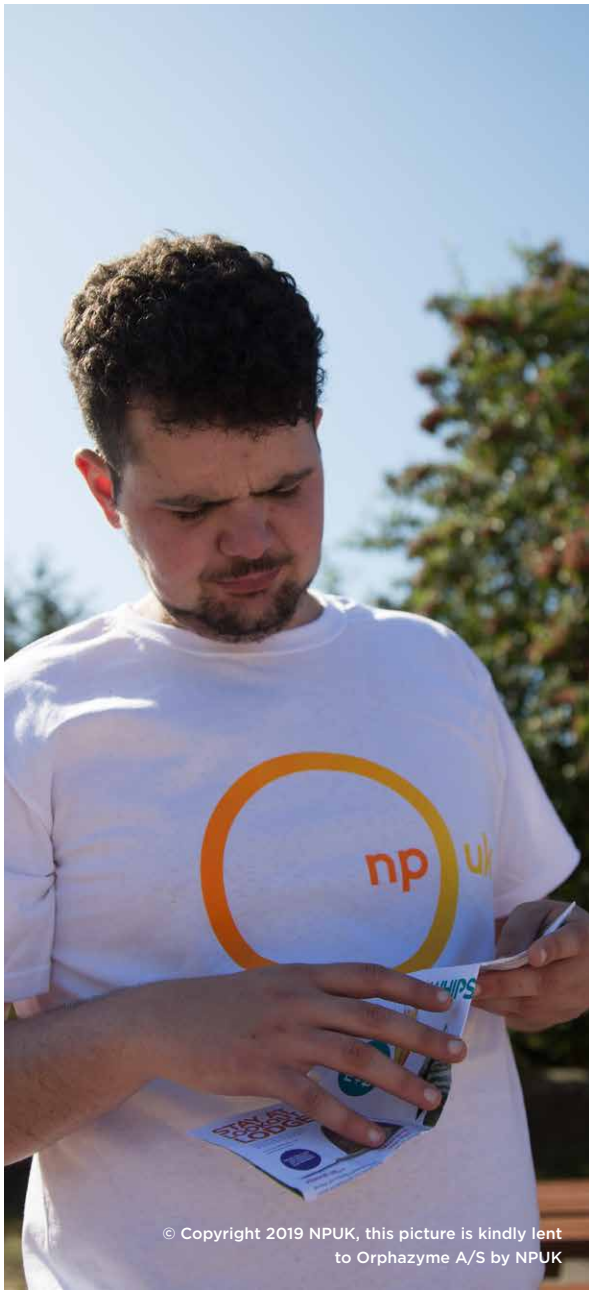
New Drug Application (NDA)
A submission to apply for marketing approval for a drug from the FDA.

Niemann-Pick disease Type C (NPC)
A rare, inherited, progressive, and often fatal neurodegenerative disease.

Orphan Drug Designation
This program provides orphan status to drugs and biologics, which are defined as those intended for the safe and effective treatment, diagnosis or prevention of rare diseases.

Rare Pediatric Disease Designation
The Rare Pediatric Disease Designation is granted by the FDA to drugs that have a potential to treat rare pediatric diseases. Orphazyme has received such a designation for arimoclomol for NPC. The designation entails the potential receipt of a so-called Priority Voucher upon marketing authorization, if certain criteria are met. The voucher can be redeemed to provide Priority Review of a subsequent NDA for a different product.

US Food and Drug Administration (FDA)
US regulatory agency responsible for ensuring the safety, efficacy and security of human and veterinary drugs, biological products and medical devices.



CORPORATE INFORMATION

COMMERCIAL BANKERS

Danske Bank
Holmens Kanal 2-12
DK-1092 Copenhagen K

Nordea
Vesterbrogade 8
DK-1620 Copenhagen

LEGAL COUNSEL

Gorrissen Federspiel, Advokatpartnerselskab
Axeltorv 2
DK-1609 Copenhagen V

INDEPENDENT AUDITORS

EY
Dirch Passers Allé 36
DK-2000 Frederiksberg

Annual report
This annual report will be available on www.orphazyme.com and printed copies are available upon request.

Annual General Meeting
The Annual General Meeting will be held on March 26, 2020 at 5.00 PM CET at:
COBIS, Ole Maaløes Vej 3,
DK-2200 Copenhagen N

2019
CONSOLIDATED FINANCIAL STATEMENTS

TABLE OF CONTENTS

FINANCIAL STATEMENTS

CONSOLIDATED STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

For the years ended December 31

(TDKK)	Note	2019	2018
Research and development expenses	2.1, 2.2	(285,413)	(196,525)
General and administrative expenses	2.3	(50,541)	(35,127)
Operating loss		**(335,954)**	**(231,652)**
Financial income	2.6	316	5
Financial expenses	2.6	(7,359)	(3,453)
Loss before tax		**(342,997)**	**(235,100)**
Income tax benefit	2.7	5,500	5,500
Net loss for the year		**(337,497)**	**(229,600)**
Items that will be reclassified subsequently to the Statement of Profit or Loss:			
Exchange difference from translation of foreign operations net of tax DKK 0		67	42
Total comprehensive loss		**(337,430)**	**(229,558)**
Loss per share, basic and diluted	4.3	(16.87)	(11.49)

CONSOLIDATED STATEMENTS OF FINANCIAL POSITION

As of December 31

ASSETS (TDKK)	Note	2019	2018
Non-current assets			
Licenses	3.1	10,539	10,744
Right-of-use assets	3.2	13,903	-
Property, plant, and equipment	3.3	3,685	1,940
Corporation tax receivable	2.7	2,750	2,750
Prepayments and deposits	3.4	1,652	2,531
Total non-currents assets		**32,529**	**17,965**
Current assets			
Corporation tax receivable	2.7	5,500	5,500
Prepayments and other receivables	3.4	19,137	23,178
Cash	3.6	123,588	394,706
Total current assets		**148,225**	**423,384**
Total assets		**180,754**	**441,349**

EQUITY AND LIABILITIES (TDKK)	Note	2019	2018
Equity			
Share capital	4.2	19,984	19,939
Share premium	4.2	924,021	924,021
Other reserves		7,982	9,112
Accumulated deficit		(899,018)	(564,823)
Total equity		**52,969**	**388,249**
Non-current liabilities			
Borrowings	3.5	51,606	-
Lease liability	3.2	9,813	-
Other non-current liabilities	3.5	378	105
Total non-current liabilities		**61,797**	**105**
Current liabilities			
Current borrowings	3.5	12,813	-
Trade payables and accruals	3.5	32,390	42,183
Other liabilities	3.5	20,785	10,812
Total current liabilities		**65,988**	**52,995**
Total equity and liabilities		**180,754**	**441,349**

CONSOLIDATED STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

(TDKK)	Notes	Share capital	Share premium	Other reserves: Foreign currency translation reserve	Other reserves: Share-based compensation – acquisition of intangible assets	Accumulated deficit	Total
Balance as of December 31, 2017		19,928	924,021	-	9,972	(338,219)	615,702
Net loss for the year						(229,600)	(229,600)
Other comprehensive loss				42		-	42
Total other comprehensive loss				42	-	(229,600)	(229,558)
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	11			(902)	891	-
Share-based compensation expense	2.5					2,105	2,105
Total transactions with owners		11	-	-	(902)	2,996	2,105
Balance as of December 31, 2018		**19,939**	**924,021**	**42**	**9,070**	**(564,823)**	**388,249**
Net loss for the year						(337,497)	(337,497)
Other comprehensive loss				67		-	67
Total other comprehensive loss				67	-	(337,497)	(337,430)
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	26			(1,197)	1,171	-
Issuance of Matching Shares, net of costs	2.5	19					19
Share-based compensation expense	2.5	-				2,131	2,131
Total transactions with owners		45	-	-	(1,197)	3,302	2,150
Balance as of December 31, 2019		**19,984**	**924,021**	**109**	**7,873**	**(899,018)**	**52,969**

See accompanying notes to these financial statements.

CONSOLIDATED STATEMENTS OF CASH FLOWS

FOR THE YEARS ENDED DECEMBER 31 (TDKK)	Note	2019	2018
Operating loss		(335,954)	(231,652)
Reversal of non-cash items:			
Equity-settled share-based compensation expense	2.5	2,549	2,105
Depreciation and amortization	3.1, 3.2, 3.3	3,803	1,366
Exchange rate adjustments		-	(491)
Change in working capital:			
Change in prepayments, deposits, and other receivables	3.4	4,920	(14,578)
Change in trade payables, accruals, and other liabilities	3.5	(2,844)	5,943
Corporation taxes received	2.7	5,500	5,500
Interest paid		(4,793)	(2,957)
Net cash used in operating activities		**(326,818)**	**(234,764)**
Investing activities			
Purchase of intangible assets	3.1	(508)	(1,603)
Purchase of property, plant, and equipment	3.3	(2,777)	(743)
Net cash used in investing activities		**(3,285)**	**(2,346)**
Financing activities			
Proceeds from borrowings	3.5	62,758	-
Repayment of lease obligations	3.2	(3,838)	-
Proceeds from issuance of shares	2.5	19	-
Net cash provided by financing activities		**58,939**	**-**
Net change in cash		(271,164)	(237,110)
Effects of changes in exchange rates		46	81
Cash at the beginning of the year		394,706	631,735
Cash at the end of the year		**123,588**	**394,706**

See accompanying notes to these financial statements.

SECTION 1

BASIS OF PREPARATION

Section 1 provides a summary of the significant accounting policies applied by the Group, Management's key accounting estimates and judgements, and new IFRS standards applicable to the Group. A detailed description of accounting policies and key accounting estimates and judgements related to specific financial statement line items is presented in each note to the relevant line item.

The consolidated financial statements of the Group have been prepared in accordance with International Financial Reporting Standards, or IFRS, as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act. The consolidated financial statements of Orphazyme A/S for the year ended December 31, 2019 were approved by the Board of Directors on February 28, 2020 and will be submitted to the shareholders of Orphazyme A/S for approval at the Annual General Meeting to be held on March 26, 2020.

In this section

1.1 CORPORATE INFORMATION

Orphazyme A/S (the "Company") is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. In April 2018, a fully-owned subsidiary, Orphazyme US, Inc., was incorporated in Massachusetts, USA (together with Orphazyme A/S, "Orphazyme" or the "Group"). Orphazyme US, Inc. will directly support the US market to establish closer relationships with the medical, patient, and financial communities.

1.2 SIGNIFICANT ACCOUNTING POLICIES

The consolidated financial statements have been prepared on a historical cost basis except for share-based compensation, which is measured at fair value.

The consolidated financial statements are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Company. Where indicated, amounts are rounded to the nearest thousand, or TDKK. The functional currency of Orphazyme US, Inc. is the US dollar (USD).

Principles of consolidation

The consolidated financial statements of the Group include the financial statements of the parent company, Orphazyme A/S (the "Parent Company") and Orphazyme US, Inc., a fully-owned subsidiary over which the Parent Company has control. A company controls an entity when the company (i) is exposed to, or has rights to, variable returns from its involvement with the entity, (ii) has power over the entity (i.e. existing rights that give it the current ability to direct the activities of the entity), and (iii) has the ability to use its power to affect the returns of the entity.

The Parent Company reassesses whether or not it controls an entity if facts and circumstances indicate that there are changes to one or more of the three elements of control. Consolidation of an entity begins when the Parent Company obtains control and ceases when the Parent Company has lost control of the entity.

Orphazyme US, Inc. has adopted the accounting policies of the Parent Company and therefore the Group's consolidated financial statements have been prepared by combining similar accounting items on a line-by-line basis. On consolidation, intercompany income and expenses, intercompany receivables, and payables, and unrealized gains and losses on transactions between the consolidated companies are eliminated.

1.2 SIGNIFICANT ACCOUNTING POLICIES (CONTINUED)

Translation of foreign currencies
Items included in the financial statements of each of the Orphazyme entities are measured using the currency of the primary economic environment in which the entity operates, or functional currency. On initial recognition, transactions denominated in foreign currencies are translated at the foreign exchange spot rate at the transaction date. For monetary assets and liabilities, differences arising between the foreign exchange spot rates at the transaction date and the date of settlement or period-end exchange rates are recognized in the Statement of Profit or Loss as financial income or financial expenses.

On consolidation, the assets and liabilities of Orphazyme US, Inc are translated from USD to DKK at the exchange rate in effect at the balance sheet date and the Statement of Profit or Loss and Other Comprehensive Income is translated from USD to DKK at the date of the underlying transaction or average exchange rate of the period if there are no significant fluctuations in exchange rate throughout the period. The exchange rate differences arising on translation for consolidation are recognized in other comprehensive income (loss).

Segment information
Although Orphazyme established a US subsidiary in 2018, the Group is managed and operated as one business unit that is reflected in the internal reporting. No separate lines of business or separate business entities have been identified with respect to any product candidate or geographical market and no segment information is currently disclosed in the Group's internal reporting. For the years ended December 31, 2019 and 2018, the Group generated no revenue and all non-current assets were located in Denmark.

1.3 KEY ACCOUNTING ESTIMATES AND JUDGEMENTS

The use of reasonable estimates and judgements is an essential part of the preparation of the consolidated financial statements. Given the uncertainties inherent in the Group's business activities, Management must make certain key accounting estimates and judgements, which affect the application of accounting policies and therefore the reported amounts of assets, liabilities, revenue, expenses, and disclosures in the consolidated financial statements. The key accounting estimates and judgements identified are those that have a significant risk of resulting in a material adjustment to the consolidated financial statements.

Management bases its estimates on historical experience, assumptions, and information currently available and deemed to be reasonable at the time the consolidated financial statements are prepared. However, actual amounts may differ from the estimated amounts as more detailed information becomes available. Estimates and assumptions are reviewed on an ongoing basis and, if necessary, changes are recognized in the period in which the estimate is revised. Management has made key accounting estimates and judgements in the following areas:

- Key estimate of research and development expenses associated with clinical trials (Note 2.1) and related prepayments (Note 3.4) and accruals (Note 3.5)
- Key estimate of inputs and assumptions used in share-based compensation valuation models (Note 2.5)
- Key estimate of the fair value of licenses (Note 3.1)
- Key estimate relating to the incremental borrowing rate to measure lease liabilities (Note 3.2)
- Key judgement regarding the recognition of deferred tax assets related to taxable losses to be carried forward (Note 2.7)
- Key judgement regarding management's assessment of the company's ability to continue as a going concern (Note 4.1)

Please refer to the specific referenced notes for further information on the key accounting estimates and judgements as well as assumptions applied.

1.4 NEW IFRS STANDARDS APPLICABLE TO THE GROUP

On January 1, 2018, the Group adopted IFRS 9, Financial Instruments, which did not have a significant impact on the consolidated financial statements. In addition, there have been amendments to IFRS 2, Share-Based Payment, which the Group adopted, but did not have any impact on the Group's consolidated financial statements. The Group has not early adopted any other standard, interpretation, or amendment that has been issued but is not yet effective. IFRS 15, Revenue from Contracts with Customers, was effective on January 1, 2018, however, as the Group does not generate revenue, the standard is not applicable at this time.

Updates to the Group's accounting policies

IFRS 16 Leases

Upon adoption of IFRS 16, the Group applied a single recognition and measurement approach for all leases except for short-term leases and leases of low-value assets. Refer to Note 3.2 Leases for the accounting policy beginning January 1, 2019. The standard provides specific transition requirements and practical expedients, which have been applied by the Group. On January 1, 2019, Orphazyme adopted IFRS 16 using the modified retrospective method. Under this method, the cumulative effect of initially applying IFRS 16 is recognized at January 1, 2019. Upon adoption, Orphazyme recognized a right-of-use asset and a lease liability based on the present value of the remaining lease payments in the amount of TDKK 13,006 for the office lease previously classified as an operating lease using a weighted average incremental borrowing rate of 3.54%. Since the application of IFRS 16, Orphazyme has recognized finance expense on the lease liability and depreciation expense on the right-of-use asset.

IFRIC Interpretation 23 Uncertainty over Income Tax Treatment

The Interpretation addresses the accounting for income taxes when tax treatments involve uncertainty that affects the application of IAS 12 Income Taxes and does not apply to taxes or levies outside the scope of IAS 12, nor does it specifically include requirements relating to interest and penalties associated with uncertain tax treatments. The Interpretation specifically addresses the following:

- Whether an entity considers uncertain tax treatments separately
- The assumptions an entity makes about the examination of tax treatments by taxation
- How an entity determines taxable profit (tax loss), tax bases, unused tax losses, unused tax credits and tax rates
- How an entity considers changes in facts and circumstances

The Group determines whether to consider each uncertain tax treatment separately or together with one or more other uncertain tax treatments and uses the approach that better predicts the resolution of the uncertainty. The Group applies significant judgement in identifying uncertainties over income tax treatments. Since the Group operates in a multinational environment, it assessed whether the Interpretation had an impact on its consolidated financial statements. Upon adoption of the Interpretation, the Group considered whether it has any uncertain tax positions, including those relating to transfer pricing. The Group determined, based on its tax compliance that it is probable that its tax treatments will be acceptable by the taxation authorities. The Interpretation did not have an impact on the consolidated financial statements.

SECTION 2

RESULT OF THE YEAR

Section 2 presents details related to Orphazyme's Statement of Profit or Loss and Other Comprehensive Income, including Research and Development expenses and General and Administrative expenses, Government Grants, Employee costs, and Share-based Compensation costs. The Group does not yet generate revenue. In addition, this section comprises Financial Income, Financial Expenses, and Income Taxes.

In this section

2.1 RESEARCH AND DEVELOPMENT EXPENSES

§ ACCOUNTING POLICIES

Research expenses comprise costs incurred during the very early stages of the drug development cycle from initial drug discovery until the drug is ready for administration to humans. The activities initially focus on identifying a single drug candidate with a profile that will support a decision to initiate development activities. Before selection of the final drug candidate, it is tested in animals to gather efficacy, toxicity and pharmacokinetic information.

Development expenses comprise costs incurred during the different phases of clinical drug development starting in phase 1, when the drug is administered to humans for the first time, through phases 2 and 3, and subsequent activities to obtain marketing authorizations, which will permit Orphazyme to eventually market and sell the drug products.

In line with industry practice, Orphazyme expenses all research costs. Development costs that do not meet the definition of an asset are also expensed as incurred. Due to regulatory and other uncertainties inherent in the development of new products, development costs do not qualify for capitalization as intangible assets until marketing approval by a regulatory authority is obtained or highly probable.

Clinical trial costs are a significant component of research and development expenses. The Company's clinical trials are performed by third-party Clinical Research Organizations (CROs) and in order to estimate the amount of costs to charge to expense Management has developed expense models for each clinical trial based on estimates and assumptions.

The clinical trials generally have three distinctive stages.

- Start-up stage: initial setting up of the trial
- Treatment stage: site and trial management during the dosing period
- Wrap-up stage: close down and reporting of the trial

For each clinical trial for which information about the actual services delivered by the CRO are not provided on a regular current basis, the Company reviews the approved budgets for the clinical trial from the original executed agreements and categorizes the individual costs according to the three stages described above. The start-up activities, which include site recruitment, regulatory applications and investigator meetings, usually are performed reasonably uniformly throughout the start-up stage and the related costs are expensed ratably over this stage, which reflects the manner in which related services are renderd by the CRO.

2.1 RESEARCH AND DEVELOPMENT EXPENSES (CONTINUED)

The start-up stage is followed by the treatment stage, during which patients are dosed with the drug under study and results are monitored and measured. The costs incurred in this stage of the trial, which comprises the major portion of the total cost of the clinical trial, is mainly driven by the number of enrolled patients undergoing treatment. The Company estimates the costs attributable to activities performed in this stage of the trial on a per-patient basis. These costs are expensed over the treatment stage as patients are enrolled and undergo treatment, as reported by the CRO. After the last patient has been treated, the trial begins to be closed down and activities are performed related to data quality assurance and analysis. These activities are performed reasonably uniformly throughout the wrap-up stage and are expensed ratably over this last stage. Other costs, such as central laboratory costs and drug supply costs, are expensed as incurred, which is typically when the service has been rendered or the goods delivered.

CROs invoice the Company upon the occurrence of predetermined milestones (such as the enrollment of patients); however, the timing of these invoices and the Company's related payments often do not correspond directly to the level of performance of contracted activities. To the extent payments are made by the Company in advance of the related activities performed by the CROs, they are included in prepayments to vendors (see Note 3.4) and expensed in accordance with the expense model discussed above. To the extent that the payments are made by the Company following the performance of the related activities, the expense is reflected as an accrual (see Note 3.5) in accordance with the expense model.

Research and development expenses include costs arising from research and clinical development activities including employee costs for research and development personnel (i.e. salaries, bonuses, employer contributions to pension schemes, share-based compensation), legal expenses related to the protection, defense and enforcement of the Company's intellectual property, as well as depreciation on right-of-use assets associated with facilities and equipment used for research and development purposes.

The following table presents research and development expenses recognized for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
External costs	216,589	152,820
Employee costs (Note 2.4)	64,167	40,281
Facility costs	1,554	2,132
Depreciation and amortization (Notes 3.1, 3.2, 3.3)	3,103	1,292
Total research and development expenses	**285,413**	**196,525**

Key estimate of research and development expenses associated with clinical trials

Accounting for clinical trial costs related to activities performed by Clinical Research Organizations (CROs) and other external vendors requires Management to make significant estimates in regard to the timing of the expense recognition of these costs. The diverse nature of services being provided by CROs, the different compensation arrangements that exist for each type of service, and the limitation in the availability of information related to when certain clinical activities are performed add complexity to the estimation of the timing of expense recognition for services rendered by CROs and other vendors in connection with clinical trials.

2.2 GOVERNMENT GRANTS

§ ACCOUNTING POLICIES

Government grants are recognized when there is reasonable assurance that the funding will be received and all underlying conditions will be fulfilled. Income from grants is recognized in the Statement of Profit or Loss as a reduction of the related expenses being reimbursed in the period when the related expenses are incurred.

Government grants comprise research funding from the Danish government and the European Union. The grants received by Orphazyme provide reimbursement for certain project-specific research and development expenses, including wages and salaries.

During the year ended December 31, 2019, Orphazyme has received TDKK 115 (2018: TDKK 2,126) in government grant funding, which was receivable as of December 31, 2018.

As of the year ended December 31, 2019, the total amount still receivable under these grants is TDKK 357 (2018: TDKK 1.237) and is classified as Current Other Receivables in the Statement of Financial Position, as all remaining funding from grants is receivable within the next year (Note 3.4).

One grant has been paid to Orphazyme in advance and income in the amount of TDKK 95 (2018: TDKK 299) related to this grant has been deferred and presented in the Statement of Financial Position as current other liabilities (Note 3.5).

All the grants received are subject to repayment clauses upon breach of conditions to maintain the terms under which the grant was awarded. Orphazyme has complied with and anticipates continuing to fully comply with all such terms.

2.3 GENERAL AND ADMINISTRATIVE EXPENSES

§ ACCOUNTING POLICIES

General and administrative expenses include salaries for administrative employees and Executive Management, remuneration to the Board of Directors, share-based compensation costs, depreciation on right-of-use assets associated with facilities not used for research and development purposes, and investor relations. In addition, we include pre-commercial activities in general and administrative expenses, such as the preparation of an Early Access Program for NPC, tradename costs, market and pricing studies and related costs.

The following table presents general and administrative expenses for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
External costs	20,326	12,471
Employee costs (Note 2.4)	25,995	15,803
Travel and related expenses	1,166	4,115
Pre-commercial activities	2,355	2,664
Depreciation (Notes 3.2 and 3.3)	699	74
Total general and administrative expenses	**50,541**	**35,127**

2.4 EMPLOYEE COSTS

§ ACCOUNTING POLICIES

Employee costs primarily comprise salaries, bonuses, social security contributions, share-based compensation, vacation and sick leave as well as pension contributions. The cost of these benefits is recognized as an expense as services are received. All employee pension plans are defined contribution plans and not defined benefit plans.

Employees are eligible to receive a discretionary bonus subject to certain predefined and individual goals as determined by the Board of Directors. Employees are also eligible to receive an extraordinary bonus at the discretion of the Board of Directors.

Executive Management consists of the Company's Chief Executive Officer and the Chief Financial Officer, also the registered management of the Company. In July 2019, Orphazyme announced that the Board of Directors appointed Kim Stratton as the new Chief Executive Officer, succeeding Anders Hinsby on October 1, 2019. In the event Orphazyme terminates the service agreement with the CEO without cause, Orphazyme is obliged to pay the CEO two times her annual salary as severance at the time of her last salary payment.

The Executive Management is eligible to receive an annual performance-based cash bonus subject to certain predefined corporate and individual goals as determined by the Board of Directors on an annual basis. A cash bonus received under the short-term incentive program may not exceed 100% of the annual fixed salary of the participants. The Executive Management is also eligible to receive an extraordinary bonus at the discretion of the Board of Directors.

The following table presents Employee Costs, including remuneration to the Board of Directors, for the years ended December 31, 2019 and 2018. Executive Management remuneration includes remuneration to Kim Stratton for the period October 1 - December 31, 2019; Anders Vadsholt for the full year; and Anders Hinsby for the full year, as he received salary and benefits for the full year 2019.

REMUNERATION TO INDIVIDUAL MEMBERS OF EXECUTIVE MANAGEMENT (TDKK)	2019	2018
Anders Hinsby (former CEO)		
Salary	2,424	1,917
Bonus	1,038	723
Share-based compensation	294	676
Other employee benefits	270	215
Total	**4,026**	**3,531**
Kim Stratton (current CEO since Oct 1, 2019)		
Salary	962	-
Bonus	1,025	-
Share-based compensation	-	-
Other employee benefits	215	-
Total	**2,202**	**-**
Anders Vadsholt (CFO)		
Salary	**1,803**	**1,411**
Bonus	**1,250**	**450**
Share-based compensation	406	463
Other employee benefits	260	161
Total	**3,719**	**2,485**
Total remuneration to the Executive Management	**9,947**	**6,016**

2.4 EMPLOYEE COSTS (CONTINUED)

(TDKK)	2019	2018
Employee costs, excluding Executive Management and Board		
Salaries	63,530	38,915
Cash bonus	5,394	3,410
Share-based compensation (Note 2.5)	1,705	1,006
Pensions	4,972	2,686
Other social security contributions	815	322
Other staff costs	766	966
Total employee costs, excluding Executive Management and Board	**77,182**	**47,305**
Executive Management remuneration		
Salaries	5,189	3,328
Cash bonus	3,313	1,173
Share-based compensation (Note 2.5)	700	1,139
Pensions	589	372
Other social security contributions	60	4
Other staff costs	96	-
Total Executive Management remuneration	**9,947**	**6,016**
Board of Directors remuneration		
Board and committee fees	2,594	2,584
Travel allowance	294	179
Share-based compensation (Note 2.5)	145	-
Total Board of Directors remuneration	**3,033**	**2,763**
Total employee costs	**90,162**	**56,084**
Recognized as follows in the Statement of Profit or Loss:		
Research and development expenses	64,167	40,281
General and administrative expenses	25,995	15,803
Total employee costs	**90,162**	**56,084**
Average number of full-time employees	**74**	**46**
Year-end number of full-time employees	**86**	**57**

Remuneration paid to members of the Board of Directors is made up of board and committee fees, a travel allowance, and share-based compensation related to the Restricted Share Units (RSUs) as described in Note 2.5. Board remuneration is recognized as general and administrative expenses in the Statement of Profit or Loss.

The following table lists Board of Directors remuneration for the years ended December 31, 2019 and 2018:

REMUNERATION TO INDIVIDUAL MEMBERS OF THE BOARD OF DIRECTORS (TDKK)	2019	2018
Georges Gemayel (Chairman of the Board)		
Board and committee fees	470	468
Travel allowance	64	47
Share-based compensation	28	-
Total	**562**	515
Bo Jesper Hansen (Deputy Chairman of the Board)		
Board and committee fees	395	394
Travel allowance	46	33
Share-based compensation	21	-
Total	**462**	427
Martin Bonde		
Board and committee fees	**259**	**258**
Travel allowance	-	-
Share-based compensation	16	-
Total	**275**	**258**
Martijn Kleijwegt		
Board and committee fees	285	284
Travel allowance	46	33
Share-based compensation	**16**	**-**
Total	**347**	**317**

2.4 EMPLOYEE COSTS (CONTINUED)

REMUNERATION TO INDIVIDUAL MEMBERS OF THE BOARD OF DIRECTORS (CONTINUED) (TDKK)	2019	2018
Rémi Droller		
Board and committee fees	270	**269**
Travel allowance	46	**33**
Share-based compensation	16	**-**
Total	**332**	**302**
Sten Verland		
Board and committee fees	309	**307**
Travel allowance	-	**-**
Share-based compensation	16	**-**
Total	**325**	**307**
Anders Hedegaard		
Board and committee fees	270	**269**
Travel allowance	46	**-**
Share-based compensation	16	**-**
Total	**332**	**269**
Catherine Moukheibir		
Board and committee fees	336	**335**
Travel allowance	46	**33**
Share-based compensation	16	**-**
Total	**398**	**368**
Total remuneration to the Board of Directors	**3,033**	**2,763**

2.5 SHARE-BASED COMPENSATION COSTS

§ ACCOUNTING POLICIES

Equity-settled awards

Shares awarded under the long-term incentive program ("LTIP") are equity-settled awards. The fair value of these awards is determined at the date of grant, resulting in a fixed fair value at grant date that is not adjusted for future changes in the fair value of the awards that may occur over the service period. The fair value of the LTIP awards has been determined using the Monte-Carlo model. Further details of the valuation models are presented below.

The fair value of equity-settled awards with service conditions and non-market performance conditions is recognized as compensation expense pro rata over the service period to the extent such awards are estimated to vest. The compensation expense is recognized together with a corresponding increase in equity over the period in which the performance and/or service conditions are fulfilled. The cumulative expense for the Group's share-based compensation awards recognized at each reporting date until the vesting date reflects the extent to which the vesting period has expired and Management's best estimate of the number of instruments that will ultimately vest. The expense or credit in the Statement of Profit or Loss for a period represents the movement in cumulative expense recognized as at the beginning and end of that period.

In the event that equity instruments are granted conditionally upon an equal number of equity instruments granted in prior periods not being exercised, they are treated as a new grant for the current period and a modification of the equity instruments granted in the prior period.

When the terms of an equity-settled award are modified, the minimum expense recognized is the grant date fair value of the unmodified award, provided that the original terms of the award are met. An additional expense, measured as at the date of modification, is recognized for any modification that increases the total fair value of the share-based payment transaction, or is otherwise beneficial to the employee. Where an award is cancelled by the entity or by the counterparty, any remaining fair value of the award is expensed immediately in the Statement of Profit or Loss.

Cash-settled awards

The phantom share-based incentive program established by the Group in June 2018 and the Restricted Share Units (RSU) awards to the board of directors are treated as cash-settled awards. A liability is recognized for the fair value of these awards, which is measured initially and at each reporting date up to and including the settlement date, with changes recognized at each reporting date. The fair value is expensed over the period until vesting date

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

Key estimate of inputs and assumptions used in share-based compensation valuation models

Estimating the fair value of the Group's share-based compensation programs requires determination of the most appropriate valuation model, which depends on the terms and conditions of the respective award. This estimate also requires making assumptions to determine the most appropriate inputs to the valuation model, including the expected life of the award, expected volatility, dividend pay-out ratio, and risk-free interest rate.

with recognition of a corresponding liability. The fair value is determined using the Monte-Carlo model, further details of which are presented below. The fair value of the cash-settled awards, which vest subject to obtaining a specified share price (i.e. market condition), is reported as compensation expense regardless of whether the share price condition is met if all other vesting conditions are met. For these awards, fair value is determined taking into account the probability of meeting the share price target. No expense is recognized for awards that do not ultimately vest.

Employees and Executive Management of the Group receive remuneration in the form of both equity-settled and cash-settled awards.

Long-term incentive program (equity settled)

In connection with the completion of the Company's initial public offering (IPO) on Nasdaq Copenhagen in November 2017, the Executive Management and Key Employees were offered to subscribe for Offer Shares ("Investment Shares") at the Offer Price for a maximum amount corresponding to approximately 15% (CMO) and 20% (CEO, CFO, and CSO) of their respective current annual base salaries.

Under the post-IPO long-term incentive program (2017 LTIP), the Executive Management as well as certain Key Employees of Orphazyme have subscribed to 14,875 ordinary shares ("Investment Shares") at the offer price of DKK 80. In April 2018, a Key Employee subscribed to 4,300 Investment Shares at the then-current market price of DKK 67.5. The Board of Directors may decide to offer other current or new employees of Orphazyme participation in the 2017 LTIP.

The participants in the 2017 LTIP may be allocated a number of shares in Orphazyme ("Performance Shares") at a price per Performance Share of DKK 1 at the end of a vesting period of four years from Orphazyme's first day of trading and official listing on Nasdaq Copenhagen. The number of Performance Shares shall be proportional to the potential increase in the price of Orphazyme's shares at the time of exercise compared to the offer price. The potential increase in the price of Orphazyme's shares will be calculated as the volume-weighted average share price as quoted on Nasdaq Copenhagen during the 10 trading days preceding the vesting date. The maximum allocation of Performance Shares will be six shares for the CEO and four shares for the other participants multiplied by the number of Investment Shares subscribed for in connection with the IPO. Performance Shares will be allocated on a linear scale with maximum allocation triggered by an 80% increase in share price, whereas no Performance Shares will be allocated if the price of Orphazyme's shares has increased 20% or less at the end of the vesting period. Among other things, vesting is also subject to the participants having maintained ownership of their Investment Shares and continued employment. Based on the number of Investment Shares subscribed for, a total maximum of 86,700 Performance Shares may be issued at the end of the vesting period.

In addition, subject to Board approval, the participants may also be allocated a number of shares in Orphazyme ("Matching Shares") at a price per Matching Share of DKK 1 in connection with the first anniversary of the subscription date of the Investment Shares. The number of Matching Shares shall be equal to the number of Investment Shares subscribed for and vesting will be subject to the participants having maintained ownership of their Investment Shares and continued employment during the one-year vesting period. Based on the number of Investment Shares subscribed for, a total of 14,875 Matching Shares fully vested at the end of the vesting period in November 2018. As of December 31, 2018, those Matching Shares had vested in full but had not yet been issued to the participants.

The remaining 4,300 Matching Shares vested in March 2019, at whichtime all 19,175 Matching Shares were issued in March 2019. Against a nominal payment of DKK 1 per share.

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

In August 2019, the Company initiated a 2019 long-term investment program (2019 LTIP) for the Executive Management and certain Key Employees with the same terms and conditions as the 2017 LTIP, i.e. Matching Shares and Performance Shares. After one year following the grant date, 31,250 matching shares will fully vest. The maximum number of performance shares that can vest in the 2019 LTIP is 125.000.

The fair value of all the LTIP awards was estimated using a Monte-Carlo simulation model at the respective grant dates, considering the terms and conditions on which the awards were granted.

The risk-free interest rate has been estimated on the basis of Danish government bonds with similar maturities. Expected volatility has been determined on the basis of the historic volatility of comparable listed companies. The company does not plan to pay out dividends in the forseeable future.

The following table presents the fair value of each program and the inputs used in the valuation models at the respective grant dates:

PROGRAM GRANT DATE	2019 LTIP Aug 2019	2017 LTIP Apr 2018	2017 LTIP Nov 2017
Fair value at the measurement date (TDKK)	6,214	714	3,895
Dividen yield (%)	-	-	-
Expected volatility (%)	51.8%	41.8%	46.6%
Risk-free interest rate (%)	(0.70%)	(0.28%)	(0.43%)
Expected life of awards (years)	3.42	3.58	3.88
Weighted average share price (DKK)	62.6	67.5	80

The weighted average remaining contractual life of the 2017 LTIP awards outstanding at December 31, 2019 was 1.88 years (2018: 2.88 years). The weighted average remaining contractual life of the 2019 LTIP awards outstanding at December 31, 2019 was 3.67. The exercise price for each LTIP award outstanding as of December 31, 2019 was DKK, 1 (2018: DKK 1).

The table below summarizes the activity related to the LTIP awards for the years ended December 31, 2019 and 2018:

(TDKK)	Executive Management	Key Employees	Total awards	Awards exercisable
Outstanding at December 31, 2017	**9,000**	**5,875**	**14,875**	**-**
Granted	-	4,300	4,300	
Exercised	-	-	-	
Expired	-	-	-	
Forfeited	-	-	-	
Outstanding at December 31, 2018	**9,000**	**10,175**	**19,175**	**14,875**
Granted	6,250	25,000	31,250	
Exercised	(9,000)	(10,175)	(19,175)	
Expired	-	-	-	
Forfeited	-	-	-	
Outstanding at December 31, 2019	**6,250**	**25,000**	**31,250**	**-**

For the year ended December 31, 2019, TDKK 2,131 (2018: TDKK 2,105) was recognized as compensation expense related to the LTIP awards, with a corresponding amount recognized in equity. Of the total expense, TDKK 700 (2018: TDKK 1,139) is attributed to the Executive Management.

Phantom share-based incentive program (cash-settled)

In June 2018, Orphazyme introduced a four-year phantom share-based incentive program (the "2018 Phantom Shares Program") for all employees other than the Executive Management and Key Employees under the LTIP.

In August 2019, Orphazyme initiated a 2019 Phantom Shares Program with the same terms and conditions as the 2018 Phantom Shares Program.

The Phantom Shares Programs are based on the share price of the Company and entitles the participants to a potential cash bonus if there has been an increase of at least 20% in Orphazyme's share price compared to the entry price at the grant date. The Phantom Shares Programs will not have any dilutive effect on the shareholders of Orphazyme as the phantom shares do not constitute or qualify for actual shares in Orphazyme.

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

The overall objectives of the Phantom Shares Programs are (i) to retain qualified employees, (ii) to create long-term incentive for the participants, and (iii) to align the interests of the employees with those of Orphazyme's shareholders. Each employee participating in the program earns the right to a certain number of phantom shares per month, depending on the employee's position. Subject to any adjustments to the Phantom Shares Programs made by the Board of Directors due to, for example, changes in Orphazyme's share capital structure or other significant events, each employee will be eligible to receive up to a total of 144 or 288 phantom shares under the program. By the end of each calendar year of the four-year program, the participants will have earned phantom shares free of charge.

The entry price per phantom share for both programs was DKK 61 and has been calculated on the basis of the volume-weighted average closing price of Orphazyme's share on Nasdaq Copenhagen during a period of 10 trading days prior to the introduction of the respective Phantom Shares Program. The phantom shares will automatically be settled in cash at the end of January 2023 for the 2018 Phantom Shares Program and at the end of January 2024 for the 2019 Phantom Shares Program by subtracting the entry price per share from the market price per share and multiplying the change by the total number of granted phantom shares, but only if Orphazyme's market price per share at that date exceeds the entry price per share by at least 20%. The market price per share will be based on the volume-weighted average closing price of Orphazyme's shares on Nasdaq Copenhagen during a period of 10 trading days prior to the settlement of the phantom shares.

The employee's cash award for each program is capped and cannot exceed a gross amount of DKK 37,500 or DKK 75,000 per employee, depending on the number of phantom shares allocated to the respective employee under the program. Based on the number of participants in the Phantom Shares Programs as of December 31, 2019 and 2018, the programs are expected to consist of up to a total of 12,750 and 8,361 phantom shares, respectively.

As of December 31, 2019 and 2018, all phantom shares granted under the Phantom Shares Program were only granted to employees of Orphazyme. No phantom shares were forfeited or expired, and none of the phantom shares were eligible for exercise.

As the Phantom Shares Programs are cash-settled, the fair value of the phantom shares granted as part of the program is estimated at each reporting date. For the year ended December 31, 2019, an aggregate amount of TDKK 273 (2018: TDKK 39) was recognized as compensation expense related to the Phantom Shares Programs, with a corresponding amount recognized as a non-current liability (Note 3.5).

The risk-free interest rate has been estimated on the basis of Danish government bonds with similar maturities. Expected volatility has been determined on the basis of the historic volatility of comparable listed companies.

The following table presents the inputs to the Monte-Carlo model used to estimate the fair values of the phantom shares as of December 31, 2019 and 2018:

Valuation date:	**December 31, 2019**		**Dec 31, 2018**
	2019 Program	**2018 Program**	**2018 Program**
Fair value at valuation date (TDKK)	347	205	73
Dividen yield (%)	-	-	-
Expected volatility (%)	57.4%	57%	52.3%
Risk-free interest rate (%)	(0.50%)	(0.63%)	(0.34%)
Expected life of awards (years)	4	3,08	4.08
Weighted average share price (DKK)	72,4	68,6	43.35

Bonus shares issued to KLSDC and UCL in connection with the license agreement

Please see Note 3.1.

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

Restricted Share Units (cash-settled)
In August 2019, Restricted Share Units (2019 RSUs) were granted to members of the Board of Directors. Participants may annually be granted a number of RSUs with a value corresponding to up to 50% of the participant's fixed annual base fee as member of the Board of Directors, not including committee membership fees. The value is calculated on the basis of the volume-weighted average share price of Orphazyme's shares as quoted on Nasdaq Copenhagen during the ten trading days preceding the grant date. The 2019 RSUs vest from the grant date to the date of the next annual general meeting, being March 26, 2020. Upon vesting, RSUs may be exercised within a period of twelve months from vesting (Exercise Period) at a price corresponding to the volume-weighted average share price during the ten trading days preceding the grant date (Exercise Price). In the event of a participant's resignation from the Board of Directors, any unvested RSUs will lapse without any rights of compensation. A decision not to be re-elected is not a resignation from the Board of Directors.

The RSUs are classified as a cash-settled program, as the Board of Directors may choose to settle any vested RSUs in cash. In such event, the cash settlement amount is based on the difference between the Exercise Price and the volume-weighted average share price as quoted on Nasdaq Copenhagen during the ten trading days preceding the first day of the Exercise Period.

The fair value of the 2019 RSUs was calculated using a Black-Scholes valuation model with the inputs shown in the following table. As the RSUs may be settled in cash, we have re-valued them as of year-end with updated inputs and recognized a cumulative share-based compensation expense in the amount of TDKK 145 and a corresponding short-term liability as of December 31, 2019. The Exercise Period is one year and for valuation purposes we have assumed exercise upon vesting, as the RSUs may be settled in cash.

As of December 31, 2019 no RSUs were forfeited or expired, and none of the RSUs were eligible for exercise.

The following table presents the inputs to the Black-Scholes model used to estimate the fair value of the 2019 RSUs at year-end:

	Dec 31, 2019
Fair value (TDKK)	232
Dividen yield (%)	-
Expected volatility (%)	44.4%
Risk-free interest rate (%)	(0.73%)
Expected life of awards (years)	0.25
Weighted average share price (DKK)	72.4

For the years ended December 31, the following amounts were recognized as share-based compensation:

(TDKK)	2019	2018
Long-Term Incentive Plans (LTIPs)	2,131	2,105
Share-based compensation included in equity	2,131	2,105
Phantom share programs	273	39
Restricted Share Units	145	-
Share-based compensation accrued as liabilities	418	39
Total share-based compensation expense recognized	**2,549**	**2,144**

2.6 FINANCIAL INCOME AND FINANCIAL EXPENSES

§ ACCOUNTING POLICIES

Financial income and expenses include interest income and expense, gains and losses due to changes in foreign exchange rates and other immaterial miscellaneous items. Beginning January 1, 2019, interest expense related to the right-of-use asset and interest expense related to the Loan Agreement are also recognized as financial expenses.

The following table presents the various items of financial income and expense recognized for the years end December 31, 2019 and 2018:

(TDKK)	2019	2018
Interest income on cash balances	316	5
Total financial income	**316**	**5**
Interest expense on Loan Agreement (Note 3.5)	3.239	-
Write-off of transaction costs for Loan Agreement tranche 2 (Note 3.5)	1.678	-
Loss on embedded call option (Note 3.5)	354	-
Interest expense on lease liabilities (Note 3.2)	351	-
Loss on lease modification (Note 3.2)	216	-
Interest expense on cash balances	1.213	2,824
Foreign currency exchange loss	229	490
Bank fees and other charges	78	139
Total financial expenses	**7.359**	**3,453**

2.7 INCOME TAXES

§ ACCOUNTING POLICIES

Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection with research and development activities under the Danish Tax Credit Regime.

Corporation tax receivable is recognized in the balance sheet as the tax benefit computed on the taxable loss for the year, adjusted for any changes to the prior year benefit due to changes in the taxable loss of prior years and for any taxes already paid or refunded.

Deferred tax is measured using the balance sheet liability method on all temporary differences between the carrying amount and the tax value of assets and liabilities, with the exception of temporary differences occurring at the time of acquisition and liabilities neither affecting the result of operation nor the taxable income.

As of December 31, 2019, there are no tax audits in process nor has management been notified of any pending tax audit.

2.7 INCOME TAXES (CONTINUED)

Key judgement regarding the recognition of the deferred tax assets related to taxable losses to be carried forward

Orphazyme is subject to income taxes in Denmark and in the U.S.A. The Company recognizes deferred income tax assets if it is probable that sufficient taxable income will be available in the future against which the temporary differences and unused tax losses can be utilized. Significant judgment is required to determine the amount of deferred tax assets that may be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Company's ability to successfully commercialize and defend its intellectual property. After consideration of these factors, Management has concluded that as regulatory approval has not yet been obtained as of December 31, 2019, the deferred income tax assets related to taxable losses carried forward do not meet the criteria for being recognized as assets in the Statement of Financial Position.

The Company's tax losses can be carried forward infinitely subject to the general rules on limited deductibility due to ownership changes. In Denmark, the Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 8,385 of taxable income plus 60% of taxable income above MDKK 8,385.

For the years ended December 31, 2019 and 2018, the Company has unrecognized net tax loss carry-forwards in the Danish entity in the amount of MDKK 425 and MDKK 280, respectively.

Management periodically evaluates positions taken in tax returns with respect to situations in which applicable tax regulations are subject to interpretation or uncertainty and establishes provisions, where appropriate. To date, there have not been any provisions established for uncertain tax positions.

The following table presents the total income tax benefit for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
Current tax benefit on net loss	75,459	51,722
Tax credit research and development expenses	5,500	5,500
Change in unrecognized deferred tax before tax credit	(74,961)	(51,850)
Permanent differences	(498)	128
Total income tax benefit for the period	**5,500**	**5,500**

The following table presents the reconciliation of the effective tax rate to the statutory corporate income tax rate in Denmark.

(TDKK)	2019	2018
Net loss before tax	(342,997)	(235,100)
Corporate income tax rate in Denmark	22%	22%
Computed income tax benefit	**75,459**	**51,722**
Tax effect of:		
Other non-deductible expenses including increased R&D deductions for tax puposes and share-based compensation	(498)	128
Deferred tax asset not recognized after tax credit	(69,461)	(46,350)
Total income tax benefit for the period	**5,500**	**5,500**

The following table presents the carrying amount of deferred tax in the Statement of Financial Position:

(TDKK)	2019	2018
Tax deductible losses	93,484	61,647
Deferred tax on intangible assets	74,050	35,887
Other temporary differences	759	738
	168,292	98,272
Deferred tax asset not recognized	168,292	98,272
Carrying amount included on statement of financial position	**-**	**-**

SECTION 3

OPERATING ASSETS AND LIABILITIES

Section 3 presents details of the operating assets and liabilities that form the basis of Orphazyme's activities, including Licenses, Right-of-Use Assets and Lease Liabilities, Property, Plant, and Equipment, Prepayments, Deposits, and Other Receivables, Financial Assets and Liabilities, Cash, and Contractual Obligations and Contingencies.

In this section

3.1 LICENSES

§ ACCOUNTING POLICIES

License rights to develop and commercialize products that are accounted for as intangible assets and acquired separately are measured on initial recognition at cost. For acquisition of intangible rights involving equity-settled share-based payment transactions, Management measures the fair value of the rights received and the corresponding increase in equity by reference to the fair value of the rights received, unless that fair value cannot be estimated reliably. If Management cannot estimate reliably the fair value of the rights received, it measures the fair value and the corresponding increase in equity by reference to the fair value of the equity instruments granted.

Following initial recognition, intangible assets are carried at cost less any accumulated amortization and accumulated impairment losses. The useful lives of intangible assets are assessed as either finite or indefinite. Intangible assets with finite lives such as license rights to develop and commercialize products are amortized over the useful economic life and assessed for impairment whenever there is an indication that the intangible asset may be impaired.

The amortization period and the amortization method for an intangible asset with a finite useful life are reviewed at least at the end of each reporting period. Changes in the expected useful life or the expected pattern of consumption of future economic benefits embodied in the asset are considered to modify the amortization period or method, as appropriate, and are treated as changes in accounting estimates. The amortization expense on intangible assets with finite lives

Key estimate of the fair value of licenses

Licenses contains an agreement entered into with the University of Kansas and University College London, in which the Company will obtain access to data and knowhow generated in the course of research in connection with the sIBM trial. Consideration for the license is to be paid out by issuing new shares to the contract partners for a value corresponding to the costs incurred during the preceding calendar year. The valuation of the license upon the execution of the agreement involves uncertainty and was estimated by Management based on the expected costs over the contract period. In addition, the estimation of the duration of a license agreement at times involves uncertainty if termination is dependent on a time limit after successful commercialization. Management has considered potential commercialization dates and will re-assess this estimate on an ongoing basis.

3.1 LICENSES (CONTINUED)

is recognized in the Statement of Profit or Loss in the expense category that is consistent with the function of the intangible assets.

CytRx Asset Purchase Agreement
In May 2011, Orphazyme entered into an Asset Purchase Agreement with the US biopharmaceutical company CytRx. Pursuant to this agreement, CytRx irrevocably sold and transferred certain preclinical and clinical data, intellectual property rights, and other assets, including contractual rights and obligations relating to a portfolio of chemical compounds, which includes arimoclomol, to Orphazyme. Under the terms of the Asset Purchase Agreement, Orphazyme agreed to make future payments to CytRx that were contingent upon the achievement of specified clinical, regulatory, and sales milestones. The Asset Purchase Agreement further includes royalty payments to be made by Orphazyme based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In August 2018, the Company made a milestone payment of USD 250,000 (TDKK 1,603) upon the enrollment of the first patient in the ALS clinical trial. The Company has capitalized this amount as an acquired license right, as Management assesses that the consideration paid reflects market expectations about the probability that future economic benefits will flow to the Company. The acquired license is not being amortized until approval of the underlying asset has been received from regulatory authorities.

License Agreement with KLSDC and UCL
In 2017, the Company entered into a license agreement with KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., ("KLSDC") and UCL Business PLC ("UCL") granting Orphazyme the right to develop and commercialize products under all data generated in the course of the on-going Phase II/III clinical trial on arimoclomol for the treatment of sIBM worldwide. The total consideration for the license is to be paid out in bonus shares to KLSDC and UCL up to an aggregate value of USD 2.5 million (DKK 15.8 million), depending on the amount of grants awarded to KLSDC and UCL for use in the trial. At the time the license agreement was executed, Management estimated the aggregate amount of the funding to be received by KLSDC and UCL to be USD 1.583 thousand (TDKK 9.972), which has been recognized as an intangible asset (License) with a corresponding increase in equity reserves (Share-based compensation-acquisition of intangible assets).

Consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. As at December 31, 2018 the aggregate costs incurred by KLSDC and UCL amounted to USD 190 thousand (TDKK 1,197), and a total of 26,060 Bonus Shares were issued to KLSDC and UCL in January 2019, based on the average 30-day closing price of Orphazyme's shares. In addition, at the time of the share issuance the equity reserve was decreased by TDKK 1,197, which represents the market value of the shares issued. See Note 4.7 for Bonus Shares issued in January 2020 in connection with the 2019 costs incurred by KLSDC and UCL.

Under the terms of the license agreement, Orphazyme shall furthermore pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme expects to generate income from such products sold for the treatment of sIBM which will exceed any royalty payments due. Orphazyme has no liabilities prior to the occurrence of future sales of products sold for the treatment of sIBM and accordingly, neither such liabilities nor contingent considerations have been recognized as part of the rights acquired.

The license is being amortized over the duration of the license agreement, which has been estimated to be approximately 14 years. Amortization expense for the years ended December 31, 2019 and 2018 amounts to TDKK 713 and TDKK 712, respectively, and is recognized within research and development expenses.

License Agreement with the University of Miami
In September 2019, the Company entered into an exclusive license agreement with the University of Miami. Pursuant to the exclusive license agreement, the Company was granted a global royalty bearing, exclusive license to use or apply the study data, know-how and patent rights generated by the University of Miami in a phase II clinical trial of arimoclomol for the treatment of ALS with the SOD1 mutation. The Company was also granted internal development use rights to the data, know-how and patent rights.

Under the terms of the exclusive license agreement, the Company made an up-front cash payment of USD 75,000 (TDKK 508) and further agreed to make future payments of certain license fees, a development milestone payment upon receiving regulatory approval for ALS and annual fees as well as a royalty of 0.75% of net sales of products sold within ALS linked to mutations in the SOD1 gene. Any annual fees will be creditable against royalty and milestone payments. Orphazyme has no liabilities prior to the occurrence of future sales of products and accordingly neither such liabilities nor contingent consideration have been recognized as part of the license agreement.

The up-front cash payment was capitalized as an acquired license right, which is not being amortized until approval of the underlying asset has been received from regulatory authorities.

3.1 LICENSES (CONTINUED)

The following table presents the cost and respective amortization of the licenses held by Orphazyme:

(TDKK)	2019
Cost at December 31, 2017	9,972
Additions	1,603
Cost at December 31, 2018	11,575
Additions	508
Cost at December 31, 2019	**12,083**
Accumulated amortization at December 31, 2017	119
Amortization expense	712
Accumulated amortization at December 31, 2018	831
Amortization expense	713
Accumulated amortization at December 31, 2019	**1,544**
Net carrying value at	
December 31, 2018	10,744
December 31, 2019	**10,539**

3.2 LEASES

§ ACCOUNTING POLICIES

The Group has applied IFRS 16 using the modified retrospective approach and therefore the comparative information has not been restated and continues to be reported under IAS 17 and IFRIC 4. The details of accounting policies under IAS 17 and IFRIC 4 are disclosed separately if they are different from those under IFRS 16.

Policy applicable before January 1, 2019

Prior to January 1, 2019, the Group classified a lease at its inception date as a finance lease or an operating lease. As lease contract where the lessor retains the significant risks and rewards associated with ownership of the asset was classified as an operating lease. A lease contract that transferred substantially all the risks and rewards associated with ownership to the Group was classified as a finance lease. The Group had not entered into any finance leases and was only party to operating leases as a lessee.

Operating lease payments were recognized as an operating expense in the Statement of Profit or Loss on a straight-line basis over the lease term. Lease payments related to facilities used for research purposes were recognized in Research and Development expenses. Lease payments related to facilities not used for research purposes were recognized in General and Administrative expenses.

Policy applicable from January 1, 2019

At contract inception, the Group assesses whether a contract is, or contains, a lease. A contract is, or contains, a lease if the contract conveys the right to control the use of an identified asset for a period of time in exchange for consideration. The Group is party to lease agreements only in which it is a lessee and not a lessor.

As a lessee, the Group applies a single recognition and measurement approach for all leases, except for short-term leases and leases of low-value assets. At inception or on reassessment of a contract that contains a lease component, the Group allocates the consideration in the contract to each lease component on the basis of their relative stand-alone prices.

The Group recognizes lease liabilities to make lease payments and right-of-use assets representing the right to use the underlying assets.

Right-of-use assets

The Group recognizes right-of-use assets at the commencement date of the lease (i.e., the date the underlying asset is available for use). Right-of-use assets are measured at cost, less any accumulated depreciation and impairment losses, and adjusted for any remeasurement of lease liabilities. The cost of right-of-use assets includes the amount of lease liabilities recognized, initial direct costs incurred, and lease payments made at or before the commencement date less any

3.2 LEASES (CONTINUED)

lease incentives received. Right-of-use assets are depreciated on a straight-line basis over the shorter of the lease term and the estimated useful life of the underlying asset. If ownership of the leased asset transfers to the Group at the end of the lease term or the cost reflects the exercise of a purchase option, depreciation is calculated using the estimated useful life of the asset, which for the operating eqipment under lease is ten years. The right-of-use assets are also subject to impairment.

Lease liabilities

At the commencement date of the lease, the Group recognizes lease liabilities measured at the present value of lease payments to be made over the lease term. The lease payments include fixed payments less any lease incentives receivable, variable lease payments that depend on an index or a rate, and amounts expected to be paid under residual value guarantees. The lease payments also include the exercise price of a purchase option reasonably certain to be exercised by the Group and payments of penalties for terminating the lease, if the lease term reflects the Group exercising the option to terminate. Variable lease payments that do not depend on an index or a rate are recognized as expenses in the period in which the event or condition that triggers the payment occurs. In calculating the present value of lease payments, the Group uses its incremental borrowing rate at the lease commencement date because the interest rate implicit in the lease is not readily determinable. After the commencement date, the amount of lease liabilities is increased to reflect the accretion of interest and reduced for the lease payments made. In addition, the carrying amount of lease liabilities is remeasured if there is a modification, a change in the lease term, a change in the lease payments (e.g., changes to future payments resulting from a change in an index or rate used to determine such lease payments) or a change in the assessment of an option to purchase the underlying asset. The Group's non-current lease liabilities are included as a separate line item on the Group's consolidated balance sheet and the current portion of lease liabilities is included in Other current liabilities.

Short-term leases and leases of low-value assets

The Group applies the short-term lease recognition exemption to its short-term leases of machinery and equipment (i.e., those leases that have a lease term of 12 months or less from the commencement date and do not contain a purchase option). It also applies the lease of low-value assets recognition exemption to leases of office equipment that are considered to be low value. Lease payments on short-term leases and leases of low-value assets are recognized as expense on a straight-line basis over the lease term.

Lease modifications

Lease modifications are accounted for at the effective date of modification, which is the date when both parties agree to the lease modification. Modifications are accounted for either as a separate lease or as a remeasurement of the initial lease. A modification is accounted for as a separate lease if both of the following conditions are met: (a) the modification increases the scope of the lease by adding the right to use one or more underlying assets; and (b) the consideration for the lease increases by an amount equivalent to the stand-alone price for the underlying asset. For a modification that is not a separate lease, the lease liability is remeasured using a discount rate determined at the effective date of the modification.

Key estimate relating to the incremental borrowing rate

The Group cannot readily determine the interest rate implicit in its leases, therefore it uses its incremental borrowing rate to measure lease liabilities. This is the rate of interest that the Group would have to pay to borrow over a similar term, and with a similar security, the funds necessary to obtain an asset of a similar value to the right-of-use asset in a similar economic environment. As there are no observable rates available for such a rate, the Group estimates its incremental borrowing rate using observable inputs, such as market interest rates, and is required to make certain entity-specific estimates.

The Group has lease contracts for its headquarters in Copenhagen and for machinery used in its operations. The lease terms range from three to five years. During 2019 the lease contract for its headquarters in Copenhagen was modified to include additional space, a true-up to market value for the lease as a whole, and an extension of the whole lease term. The modification was accounted for as a change in the scope of the existing lease and therefore the initial lease was remeasured on the effective date of the modification at the weighted average incremental borrowing rate of 5,28%. The effect on the right-of-use assets, lease liabilities and the Statement of Profit or Loss is disclosed in the tables below.

3.2 LEASES (CONTINUED)

The following table presents the carrying amounts of right-of-use assets recognized and the movements during the period:

(TDKK)	Office buildings	Operating equipment	Total
As at January 1, 2019	13,006	-	13,006
Additions	-	4,008	4,008
Depreciation expense	(1,858)	(200)	(2,058)
Modifications	(1,054)	-	(1,054)
As at December 31, 2019	**10,095**	**3,808**	**13,903**

The following table presents the carrying amounts of lease liabilities and the movements during the period:

(TDKK)	2019
As at January 1, 2019	13,006
Additions	4,008
Accretion of interest	351
Payments	(3,838)
Modifications	(838)
As at December 31, 2019	**12,689**
Current	2,876
Non-current	9,813

The maturity analysis of lease liabilities is disclosed in Note 3.5

The following are the amounts recognized in Statement of Profit or Loss:

(TDKK)	2019
Depreciation expense of right-of-use assets (R&D)	1,847
Depreciation expense of right-of-use assets (G&A)	211
Interest expense on lease liabilities	351
Loss on lease modification	216
Total amount recognized in profit or loss	**2,625**

3.3 PROPERTY, PLANT, AND EQUIPMENT

§ ACCOUNTING POLICIES

Property, plant, and equipment includes IT, lab and other equipment, furniture and leasehold improvements that are measured at cost less accumulated depreciation and impairment losses. Cost includes the acquisition price and costs directly related to the acquisition until the time the asset is ready for use. The residual value of equipment is not material. Depreciation is calculated on a straight-line basis over the expected useful life of the asset, being 3-5 for equipment and furniture. Leasehold improvements are depreciated over the shorter of the useful life of the improvement or the remaining lease term. The useful life of assets and method of depreciation are reviewed by management at least each year-end or more often based on changes in facts and circumstances. Changes in useful lives or residual values are adjusted prospectively as changes in accounting estimates. In addition, the Company has fully depreciated equipment still in use.

Property, plant, and equipment is required to be tested for impairment when there are impairment indicators present. Impairment tests are conducted at the individual asset level, or at the lowest level for which separately identifiable cash flows for groups of assets exist. Impaired assets or asset groups are written down to their recoverable amount, which is the higher of the value in use and the net realizable value of the asset or asset group, with impairment charges allocated proportionately to the assets within the impaired asset group.

The following table presents the Company's Property, plant and equipment as of the years presented:

(TDKK)	Furniture and equipment	Leasehold improvements	Total
Cost at December 31, 2017	3,732	346	4,078
Additions	687	56	743
Disposals	-	-	-
Cost at December 31, 2018	4,419	402	4,821
Additions	1,113	1,664	2,777
Cost at December 31, 2019	**5,532**	**2,066**	**7,598**
Accumulated depreciation at December 31, 2017	2,183	44	2,227
Depreciation expense	578	76	654
Accumulated depreciation at December 31, 2018	2,761	120	2,881
Depreciation expense	853	180	1,033
Accumulated depreciation at December 31, 2019	**3,614**	**300**	**39,14**
Net carrying value at			
December 31, 2018	1,658	282	1,940
December 31, 2019	**1,918**	**1,766**	**3,685**

There has been no impairment of property, plant and equipment for the years ended December 31, 2019 and 2018. Depreciation expense is included within operating loss as follows:

(TDKK)	2019	2018
Research and development expenses	544	580
General and administrative expenses	489	74
Total depreciation expense	**1,033**	**654**

3.4 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

§ ACCOUNTING POLICIES

Prepayments
Prepayments include advance payments made to vendors that will be incurred and expensed in subsequent financial reporting periods. When the period for full expense recognition is longer than one year from the balance sheet date, the portion to be expensed subsequent to one year is classified as non-current.

Deposits
Deposits include advance payments made to vendors to be settled upon completion of the underlying contract. When the contract term is longer than one year from the balance sheet date, the deposit is classified as non-current.

Other receivables
Other receivables include current and non-current amounts due to the Company.

Sales tax
Expenses and assets are recognized net of the amount of sales tax, except:

- When the sales tax incurred on a purchase of assets or services is not recoverable from the taxation authority, in which case, the sales tax is recognized as part of the cost of acquisition of the asset or as part of the expense item, as applicable
- When receivables and payables are stated with the amount of sales tax included

The net amount of sales tax recoverable from, or payable to, the taxation authority is included as part of receivables or payables in the Statement of Financial Position.

Key estimate of prepayments related to clinical trial development costs
As explained in Note 2.1, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. In particular, certain CROs and vendors are paid upfront in connection with clinical activities and Management is required to estimate the timing of the prepayment release to expense. This expense for the year is estimated by using an expense model, as described in Note 2.1.

The following items comprised non-current prepayments and deposits as of December 31, 2019 and 2018:

(TDKK)	2019	2018
Deposits with vendors	295	1,266
Prepayments to vendors	465	633
Leasehold deposit	892	632
Total non-current prepayments and deposits	**1,652**	**2,531**

Non-current prepayments and deposits mainly includes a deposit with a CRO for advance payment of pass-through costs in connection with a clinical trial, prepaid insurance, and the lease deposit on our headquarters in Copenhagen.

Current prepayments and other receivables are specified below:

(TDKK)	2019	2018
Prepayments to vendors	13,355	14,233
Grant income receivable	357	1,237
VAT receivable, net	2,521	1,468
Foreign VAT receivable	1,304	6,116
Other current receivables	1,600	124
Total current prepayments and other receivables	**19,137**	**23,178**

Current prepayments to vendors includes prepayments made to CROs for clinical trial costs of TDKK 5,020 (2018: TDKK 8,572).

3.5 FINANCIAL ASSETS AND LIABILITIES

§ ACCOUNTING POLICIES

Financial assets

Initial recognition and measurement

Financial assets that meet certain criteria are classified at initial recognition as subsequently measured at amortized cost, fair value through other comprehensive income (OCI), or fair value through profit or loss. The Group does not hold any financial assets meeting these classification criteria except cash and certain types of other receivables. Generally, the Company's financial assets are available to support current operations and amounts expected to be realized within the next twelve months are classified in the Statement of Financial Position as current assets.

The Group's financial assets are recognized initially at fair value plus, in the case of financial assets not carried at fair value through profit and loss, transaction costs that are attributable to the acquisition of the financial asset, if any. Financial instruments recognized at fair value are allocated to one of the following valuation hierarchy levels:

- Level 1: Quoted (unadjusted) prices in active markets for identical assets or liabilities.
- Level 2: Other techniques for which all inputs that have a significant effect on the recorded fair value are observable, either directly or indirectly.
- Level 3: Techniques that use inputs that have a significant effect on the recorded fair value that are not based on observable market data.

Subsequent measurement

Historically, the Group's receivables are due within a twelvemonth period and therefore the impact of using the effective interest rate method on the Group's financial statements has been immaterial.

Financial asset impairment

The Group assesses at the end of each reporting period whether there has been objective evidence that a financial asset may be impaired. Impairment losses are recognized if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. The Group did not assess an impairment of a financial asset for either of the years ended December 31, 2019 or 2018.

Financial liabilities

Borrowings

Financial liabilities, including borrowings, are initially measured at fair value less transaction costs incurred. Subsequently, borrowings are measured at amortized cost. Amortized cost is calculated as original cost less instalments plus/less the accumulated amortization of the difference between cost and nominal value, so that the effective interest rate is recognized in the income statement over the loan period. Financial liabilities are derecognized when settled.

The portion of the debt maturing after one year is presented as non-current debt and the remainder as current debt.

Trade payables and accruals

Trade payables and accruals relate to the Group's purchase of products and services from various vendors in the normal course of business.

Other liabilities

Other payables are measured at net realizable value. The amount payable to employees for the Phantom Shares Program (Note 2.5) is classified as non-current and is measured at fair value, at Level 2 in the fair value hierarchy.

Key estimate of accruals related to clinical trial development costs

As explained in Note 2.1, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. As described in Note 2.1, Management uses an expense model to estimate the timing of expenses recognition in each period and related accruals at the end of the year.

The Group's financial assets include mainly cash (Note 3.5). The Group has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements as discussed herein. Financial assets are not acquired for trading or speculative purposes, nor has the Group placed any assets as security for loans at either December 31, 2019 or 2018.

In August 2019, Orphazyme entered into a structured debt facility ("Loan Agreement") with Kreos Capital to secure funding of EUR 9 million (Tranche 1") to be repaid over forty-two months ("Loan Term"), with the first twelve months requiring interest only payments at nominal annual fixed interest rate of 9.75% and the remaining thirty months requiring equal installments comprising principal and interest. Early repayment of the borrowed amounts may be made in whole but not in part, with the repayment amount being equal to the principal outstanding plus the sum of all the interest repayments that would have been paid throughout the remainder of the loan discounted at an annual rate of 4.0%.

In addition, the lender may, at any time in its sole discretion in eight years, depending on certain events defined in the Loan Agreement, notify the Company that a Facilitation Fee is due and payable ("Notification").

3.5 FINANCIAL ASSETS AND LIABILITIES (CONTINUED)

The Facilitation Fee is an amount equal to the greater of (i) 10% of the aggregate amount of the amount borrowed and (ii) the percentage increase in the Company's share price between the 30-day volume-weighted average share price on the date of the Loan Agreement and the closing share price on the day immediately preceding the date of the notification applied to the aggregate amount of amounts borrowed. The variability arising from the change in Orphazyme's share price is not closely related to the host debt instrument characterized mainly by interest rate and credit risk. Therefore, the embedded equity-linked amount is separated from the host debt instrument and accounted for as an embedded written call option at fair value through profit and loss.

Fair value on inception of the Loan Agreement is included as part of the transaction costs. The call option is measured at fair value at level 2 in the fair value hierarchy.

The written call option is measured at fair value using a Black-Scholes option valuation model. In measuring the fair value, various observable and unobservable inputs are required. Observable input mainly relates to the market price of Orphazyme's shares, and risk-free interest rate. Unobservable inputs mainly relate to the expected volatility of Orphazyme's share price and the term. The table below shows the inputs used in the valuation of the call option and the estimated fair value at the date of the Loan Agreement and at December 31, 2019.

The change in fair value of the call option is recognized as a finance expense in the statement of profit or loss. During the year ended December 31, 2019, the company recognized a loss of TDKK 354.

The structured debt facility included a potential second tranche available to Orphazyme, however as of December 31, 2019 conditions allowing for the draw down of the second tranche were not met and it expired unused. In connection with the draw down of Tranche 1, Orphazyme incurred transaction costs in the amount of TEUR 450 (TDKK 3,356). As the transaction costs secured a potential financing of two tranches, half of the transaction costs TEUR 225 (TDKK 1,678) are being amortized with the first tranche and upon expiration of the second tranche, the other half of the transaction costs were written off as finance expense in the statement of profit or loss (Note 2.6).

As part of the closing of the Loan Agreement, Orphazyme made a payment in the amount of TEUR 337 (TDKK 2,511) as a deposit for the last cash payment to be made on the borrowing ("Advance Payment").

The total liability for the Loan Agreement is being amortized net of the transaction costs, the Facilitation Fee and the call option; and it is being presented net of the Advance Payment.

As of the years ended December 31, 2019 and 2018, the Group's financial liabilities comprise the following

As of the year ended December 31, 2019, Accruals includes an amount of TDKK 13,286 (2018: TDKK 13,376) for clinical trial costs.

CALL OPTION ON FACILITATION FEE	Dec 2019	Sep 2019
Fair value of call option	1,595	1,242
Dividend yield (%)	-	-
Expected volatility (%)	57%	53.3%
Risk-free interest rate (%)	(0.63%)	(0.58%)
Expected life (years)	3.2	3.5
Share price (DKK)	72.4	(63.5)

(TDKK)	2019	2018
Borrowings	62,824	-
Lease liabilities (Note 3.2)	12,689	-
Trade payables	1,093	18,090
Accruals	31,297	24,093
Total liabilities measured at amortized cost	**107,903**	**42,183**

3.5 FINANCIAL ASSETS AND LIABILITIES (CONTINUED)

Maturities of financial liabilities

The table below presents the Group's financial liabilities by relevant maturity groupings based on their contractual maturities for all non-derivative financial liabilities and derivative financial instruments for which the contractual maturities are essential for an understanding of the timing of the cash flows.

As the Facilitation Fee is due upon demand, it is shown as current Borrowings under non-derivatives. The call option on the Facilitation Fee is shown as current under derivatives.

The amounts disclosed in the table are the contractual undiscounted cash flows. Balances due within 12 months equal their carrying balances as the impact of discounting is not significant.

(TDKK)	Less than 6 months	6-12 months	Between 1 and 2 years	Between 2 and 5 years	Total contractual cash flows	Carrying amount
Non-derivatives						
Trade payables and accruals	32,390	-	-	-	32,390	32,390
Borrowings	9,997	11,147	30,165	34,695	86,004	62,824
Lease liabilties	1,672	1,672	3,344	7,405	14,093	12,689
Total non-derivatives	**44,058**	**12,819**	**33,509**	**42,100**	**132,486**	**107,903**
Derivatives (Borrowings)	1,595				1,595	1,595
Total derivatives	**1,595**				**1,595**	**1,595**

As of the years ended December 31, 2019 and 2018, total current other liabilities is comprised as follows:

(TDKK)	2019	2018
Deferred grant income	95	299
Remuneration to the Board of Directors	1,535	1,836
Lease liability	2,876	-
Payroll and employee-related costs	16,277	8,677
Total current other liabilities	**20,785**	**10,812**

Certain amounts included in prior year trade payables and accruals and other liabilities have been reclassified for consistency with current year presentation.

In addition, the Group has the following total other non-current liabilities as of the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
Accrual for milestone payment to vendor	65	66
Phantom shares liability to employees	313	39
Total non-current other liabilities	**378**	**105**

3.6 CASH

§ ACCOUNTING POLICIES

Cash includes cash on hand and in banks that is subject to the risk of changes in value (Note 4.4).

Statement of cash flows

The statement of cash flows is presented using the indirect method and shows cash flows resulting from operating activities, investing activities, financing activities, and the Group's cash at the beginning and end of the year, including any effects of exchange rate changes.

Cash flows used in operating activities converts items in the Statement of Profit or Loss from the accrual basis of accounting to the cash basis of accounting. Non-cash items such as foreign exchange gains and losses, depreciation, amortization, and changes in working capital are reversed from the net loss for the year and actual cash receipts and payments are included.

Cash flows from investing activities shows payments related primarily to the purchase of licenses and property, plant, and equipment.

Cash flows from financing activities shows proceeds from share issuance, borrowings, net of transaction costs, and lease payments.

As of the years ended December 31, 2019 and 2018, the Group's cash balance denominated in foreign currencies was as follows:

(TDKK)	2019	2018
DKK	89,155	392,196
EUR	20,083	596
USD	14,309	1,886
GBP	41	28
Total cash	**123,588**	**394,706**

3.7 COMMITMENTS AND CONTINGENCIES

Contractual obligations

The Group has entered into an operating lease for its headquarters location in Denmark. The lease component portion is accounted for under IFRS 16 (see Note 3.2) and the non-lease component portion, which consists of basic services and maintenance, has a non-cancellable lease term of six months. At December 31, 2019, the non-lease component established a contractual commitment of TDKK 494 (2018: DKK 1,333). In addition, the Group has contractual obligations related to contracts with CROs and other vendors for research and development activities that have been initiated and are non-cancelable as of December 31, 2019. These establish contractual commitments of approximately DKK 178 million (2018: DKK 178 million).

Commitments

In connection with a loan agreement in the amount up to USD 18,000,000 entered into on 27 August 2019 with Kreos Capital VI (UK) Ltd., the Company has granted security in favour of Kreos Capital VI (UK) Ltd. over (i) certain of its assets, including its intellectual property rights, pursuant to floating charge agreement registered with the Danish personal register in the initial principal amount of USD 9,000,000, (ii) its patents registered in Germany, UK and the US pursuant to a patent pledge agreement and (iii) its shares in its US subsidiary, Orphazyme US, Inc. Furthermore, Orphazyme US, Inc. has granted in favour of Kreos Capital VI (UK) Ltd. (i) a guarantee for the Company's obligations under the loan agreement pursuant to a guaranty agreement and (ii) security over certain of its assets, including its intellectual property rights, pursuant to US law security agreement.

Contingent Assets and Contingent Liabilities

The Group will recognize assets or liabilities in the future that are dependent on achieving certain milestones that may or may not be in the Group's control.

Under the terms of the asset purchase agreement with CytRx described in Note 3.1, Orphazyme agreed to make future royalty payments to CytRx based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In addition, under the terms of license agreements with KLSDC and UCL and with the University of Miami as described in Note 3.1, Orphazyme shall pay royalties of net sales as specified in the respective agreements.

Orphazyme expects to generate income from any products sold, which will exceed royalty payments due to third parties. Orphazyme has no obligation to pay royalties prior to the occurrence of the sale of products and accordingly, neither a liability for the royalties to be paid nor contingent consideration to be received as part of the rights acquired has been recognized.

3.7 COMMITMENTS AND CONTINGENCIES (CONTINUED)

The U.S. FDA has granted Orphazyme a rare pediatric disease designation to arimoclomol as a treatment for Niemann-Pick disease Type C (NPC). Under the FDA's rare pediatric disease priority review voucher program, upon the approval of a new drug application for the treatment of a rare pediatric disease, the sponsor of such application would be eligible for a rare pediatric disease priority review voucher that can be used to obtain priority review for a subsequent new drug application. However, receiving a rare pediatric disease designation for arimiclomol as a treatment for NPC does not automatically mean that the Company will receive a priority review voucher as priority review voucher is only awarded following approval by the FDA of arimoclomol as a treatment for NPC. If a priority review voucher is granted, the Company may use the voucher for its own FDA approval processes or decide to sell the voucher to other biotech or pharmaceutical companies. There is no established market for priority review vouchers and disclosed sales prices may not be indicative of the current value of vouchers, which may also fluctuate significantly. The term of the rare pediatric disease priority review vouchers program expires after September 2020 and may not be renewed or may be amended or cancelled prior to such expiry. Hence, it may be unavailable to the Company even if it meets all of the requirements. Further, the potential award of a voucher would trigger an obligation to market the relevant rare pediatric disease product within one year from FDA approval or the FDA may revoke the voucher. Finally, a voucher award subjects the Company to post marketing reporting obligations to the FDA.

SECTION 4

OTHER DISCLOSURES

Section 4 presents details of other disclosures relevant to the consolidated financial statements of the Group, including Capital Management, Equity, Earnings per Share, Financial Risk, Related Parties, Fees to Statutory Auditors, and Significant Events after the Reporting Period.

In this section

4.1 CAPITAL MANAGEMENT

For the purpose of the Group's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Group. The primary objective of the Group's capital management is to maximize shareholder value while limiting the financial risk. The Board of Directors' policy is to maintain a strong capital base in order to maintain investor, creditor and market confidence, and a continuous advancement of the Group's intellectual property, product pipeline, and business.

Cash and financial assets are monitored on a regular basis by Management and the Board of Directors in assessing current and long-term capital needs. As of December 31, 2019, the Group held cash totaling TDKK 123,588 (2018: TDKK 394,706) and that will not be sufficient to provide adequate funding to allow the Group to meet its planned operating activities. Before year-end Management commenced measures to secure adequate funding. On February 7, 2020 Orphazyme completed an offering of 7,032,937 shares in a directed issue and private placement and raised approximately DKK 745 million (see Note 4.7). The transaction is expected to cover Orphazyme's financing needs well into 2021.

4.2 EQUITY

The following table summarizes the Company's share activity:

	Ordinary shares
December 31, 2017	**19,928,184**
Issuance of bonus shares as part of license agreement (Note 3.1)	11,380
December 31, 2018	**19,939,564**
Issuance of bonus shares as part of license agreement	**26,060**
Issuance of Matching Shares (Note 2.5)	**19,175**
December 31, 2019	**19,984,799**

The Company has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

In January 2018, the Company issued 11,380 bonus shares ("2018 Bonus Shares") using free reserves of the Company to KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc. ("KLSDC"), and UCL Business PLC ("UCL") under the terms of the license agreement entered into in October 2017 (Note 3.1).

In January 2019, the Company issued 26.060 bonus shares ("2019 Bonus Shares") to KLSDC and UCL under the terms of the same license agreement mentioned above.

In March 2019, the Company issued 19,175 Matching Shares to participants in the 2017 LTIP (see Note 2.5)

Following this share capital increase, the total nominal share capital of the Company as of December 31, 2019 was DKK 19,984,779, divided into 19,984,779 ordinary shares, respectively, each with a nominal value of DKK 1.

4.3 LOSS PER SHARE

Basic loss per share for the year is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the year. The diluted loss per share is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the period increased by the dilutive effect of the assumed issuance of outstanding share-based awards. As a result of the Group incurring losses for each of the years ended December 31, 2019 and 2018, the potential shares issuable related to outstanding share-based awards have been excluded from the calculation of diluted per share amounts, as the effect of such shares is anti-dilutive.

In January 2019 and in January 2020, the Company issued 26.060 bonus shares ("2019 Bonus Shares") and 20,650 bonus shares ("2020 Bonus Shares") respectirely to KLSDC and UCL under the terms of the license agreement entered into in October 2017 with the parties (Note 3.1).

Basic and diluted loss per share for the years presented have been adjusted retrospectively to include the 2019 Bonus Shares and the 2020 Bonus Shares in the number of weighted average shares outstanding for the years ended December 31, 2019 and 2018. This results in the comparative figure for 2018 being updated accordingly.

The following reflects the net loss attributable to shareholders and share data used in the basic and diluted earnings/(loss) per share computations for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
Net loss for the year (TDKK)	(337,497)	(229,600)
Weighted-average shares outstanding	20,002,139	19,986,274
Loss per share	**(16.87)**	**(11.49)**

4.4 FINANCIAL RISKS

The Group's activities expose it to a number of financial risks whereby future events, which can be outside the control of the Group, could have a material effect on its financial position and results of operations. The known risks include foreign currency, interest and credit risk and there could be other risks currently unknown to Management. The Group has not historically hedged its financial risks.

Liquidity Risk

At December 31, 2019, the Group's liquidity risk was assessed to be high. Management continuously assesses the Group's capital structure in order to evaluate whether its liquidity reserves allow it to achieve its business objectives. At December 31, 2019, the available liquidity reserves were assessed not to be sufficient to allow it to achieve its business objectives. Before year-end Management commenced measures to secure adequate funding. On February 7, 2020 Orphazyme completed an offering of 7,032,937 shares in a directed issue and private placement and raised approximately DKK 745 million (see Note 4.7).

Foreign Currency Risk

The Group's foreign currency risk is assessed to be high. The Group's functional currency is the DKK and it conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of the DKK against the EUR, the USD and/or the GBP will expose the Group to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material.

Interest Rate Risk

The Group's interest rate risk is assessed to be low. The Group has a borrowing on which it incurs a fixed rate of interest (see Note 3.5). In addition, due to the current interest level in Denmark, the Group incurs negative interest on bank deposits.

Credit Risk

The Group's credit risk is assessed to be low. The Group's credit risk is associated with cash held in banks. The Company does not trade financial assets for speculative purposes and invests with the objective of preserving capital. The Company's cash is held primarily at two banks in Denmark with Moody's long-term credit ratings exceeding of A1.

The Group has prepared a sensitivity analysis in order to assess the potential impact on the Group's net loss for possible fluctuations in the EUR and USD exchange rates against the DKK and the impact for the possible fluctuations in the interest rate on bank deposits in Denmark and in the USA. The methods and assumptions used are consistent with prior year and consider increases and decreases in the Group's three main currencies, as well as reasonable fluctuations in the interest rate on its bank deposits. Based on these analyses, if interest rates on our cash deposits would have fluctuated by +/- 1%, the impact on the Group's net loss for the year ended December 31 ,2019 would have been approximately TDKK 8 (2018: TDKK 20). The impact of currency fluctuations on the Group's net loss is shown in the table below:

CURRENCY	Currency fluctuation	Effect 2019 TDKK	Effect 2018 TDKK
EUR	+/- 2%	503	713
USD	+/-10%	21	1,278
GBP	+/-10%	461	218

4.5 RELATED PARTIES

Orphazyme A/S, incorporated in Denmark, is the ultimate parent company of the Group, which wholly owns Orphazyme US, Inc. These two entities are considered related parties. Orphazyme A/S is not ultimately controlled by any of its investors. Major investors owning more than 10% of the Company are considered related parties.

In July 2019, Orphazyme announced that the Board of Directors appointed Kim Stratton as the new Chief Executive Officer of Orphazyme, succeeding Anders Hinsby. Kim started her position on October 1, 2019.

For the years ended December 31, 2019 and 2018, the following related party transactions were identified:

- Remuneration to Executive Management (Note 2.4)
- Remuneration to the Board of Directors (Note 2.4)
- Participation of Executive Management in the 2017 LTIP and the 2019 LTIP (Note 2.5)
- Participation of the Board members in the RSU, program (Note 2.5)

As of December 31, 2019 and 2018, the Company did not have any amounts receivable from related parties and therefore has not recorded any impairment. The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the Board of Directors or Executive Management.

During 2019 the board members have been granted RSUs, as disclosed in Note 2.5.

For the years ended December 31, 2019 and 2018, Executive Management and members of the Board of Directors had the following shareholding in Orphazyme A/S:

	Number of shares owned 2019	Number of shares owned 2018	Investment shares (LTIP) 2019	Investment shares (LTIP) 2018
Kim Stratton	-	-	-	-
Anders Hinsby	209,596	204,596	-	5,000
Anders Vadsholt	132,595	127,806	6,250	4,000

MEMBERS OF THE BOARD OF DIRECTORS:	Number of shares 2019	Number of shares 2018	Number of RSUs 2019
Georges Gemayel	97,358	87,758	3,451
Bo Jesper Hansen	100,545	79,945	2,689
Martijn Kleijwegt	-	-	1,927
Martin Bonde	46,009	46,009	1,927
Rémi Droller	-	-	1,927
Sten Verland	-	-	1,927
Anders Hedegaard	13,750	6,250	1,927
Catherine Moukheibir	7,980	7,980	1,927

4.6 FEES TO STATUTORY AUDITORS

The following table presents the fees to our statutory auditors, Ernst & Young Godkendt Revisionspartnerselskab, recognized in general and administrative expenses in the Statement of Profit or Loss for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
Audit services	2,244	320
Audit-related services	882	156
Other assistance	-	50
Total fees to statutory auditors	3,126	526

4.7 SIGNIFICANT EVENTS AFTER THE REPORTING PERIOD

In January 2020, Orphazyme established a wholly-owned subsidiary in Zug, Switzerland to facilitate the global launch of the Group's products once regulatory approval has been obtained.

As described in Note 3.1, as part of the license agreement with KLSDC and UCL, consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. As at December 31, 2019 the aggregate costs incurred by KLSDC and UCL amounted to USD 332 thousand (TDKK 2,219), and a total of 20,650 Bonus Shares ("2020 Bonus Shares") were issued to KLSDC and UCL in January 2020, based on the average 30-day closing price of Orphazyme's shares. At the time of the share issuance the equity reserve was decreased by TDKK 2,094, which represents the market value of the shares issued.

On February 7, 2020 Orphazyme completed an offering of 7,032,937 shares in a directed issue and private placement and raised approximately DKK 745 million. In gross proceeds the directed issue and private placement is expected to cover the Company's financing needs until well into 2021, and support in particular the imminent US and European filings for approval of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC), as well as the preparations for commercial launch.

The transaction consisted of a directed issue and private placement of up to 3,961,264 new shares of a nominal value of DKK 1 each (the "New Shares") and private placement of up to 3,071,673 existing shares of a nominal value of DKK 1 each (the "Existing Shares" and together with the New Shares, the "Offer Shares") at an offer price of DKK 106 per Offer Share, as determined by the Board of Directors of the Company through a book-building process (the "Offering"). The New Shares will be issued without pre-emption rights for existing shareholders.

The offering of Existing Shares is facilitated by a share loan from Novo Holdings A/S and Orpha Pooling B.V. (the "Lending Shareholders") to the Company pursuant to a stock lending and subscription agreement with an obligation for the Company to redeliver new shares of an equivalent number as the Existing Shares borrowed by the Company from each of the Lending Shareholders (the "Replacement Shares"), which will be issued without pre-emption rights for existing shareholders. The Lending Shareholders do not participate in the Offering, and are only facilitating the loan of the Lending Shares for purposes of the Company's offering of Existing Shares in the Offering.

After settlement of the capital increases, the share capital of the Company consists of 27,038,386 shares with a nominal value of DKK 1 each.

2019
PARENT COMPANY FINANCIAL STATEMENTS

STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

PARENT COMPANY

FOR THE YEARS ENDED DECEMBER 31 (TDKK)	Note	2019	2018
Research and development expenses	2.1*	(285,413)	(196,525)
General and administrative expenses	2.1	(49,682)	(35,127)
Operating loss		**(335,095)**	**(231,652)**
Financial income	2.6*	316	5
Financial expenses	2.6*	(7,359)	(3,453)
Loss before tax		**(342,138)**	**(235,100)**
Income tax benefit	2.7*	5,500	5,500
Net loss for the period		**(336,638)**	**(229,600)**
Items that will be reclassified subsequently to the Statement of Profit or Loss:			
Exchange difference from translation of foreign operation, net of tax DKK 0		-	(51)
Total comprehensive loss		**(336,638)**	**(229,651)**
Loss per share, basic and diluted		(16,35)	(11.49)

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF FINANCIAL POSITION

PARENT COMPANY

AS OF DECEMBER 31 (TDKK)	Note	2019	2018
ASSETS			
Non-current assets			
Right of use assets		13,903	-
Licenses	3.1*	10,539	10,744
Property, plant, and equipment	3.3*	3,685	1,940
Corporation tax receivable	2.7*	2,750	2,750
Prepayments and deposits	3.4*	1,652	2,531
Investment in subsidiary	2.4	-	1,207
Total non-currents assets		**32,529**	**19,172**
Current assets			
Corporation tax receivable	2.7*	5,500	5,500
Prepayments and other receivables	2.5	18,957	23,081
Cash	2.7	123,251	393,123
Total current assets		**147,708**	**421,704**
TOTAL ASSETS		**180,237**	**440,876**

EQUITY AND LIABILITIES	Note	2019	2018
Equity			
Share capital	4.2*	19,984	19,939
Share premium	4.2*	924,021	924,021
Other reserves		7,822	9,019
Accumulated deficit		(898,159)	(564,823)
Total equity		**53,668**	**388,156**
Non-current liabilities			
Lease liabilities		9,813	-
Other non-current liabilities	3.5*	378	105
Long term debt	2.5	51,611	826
Total non-current liabilities		**61,802**	**931**
Current liabilities			
Current borrowings	2.6	12,808	-
Trade payables and accruals	2.6	31,173	40,977
Other liabilities	2.6	20,785	10,812
Total current liabilities		**64,766**	**51,789**
TOTAL EQUITY AND LIABILITIES		**180,237**	**440,876**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

PARENT COMPANY

(TDKK)	Note	Share capital	Share premium	Foreign currency translation reserve	Share-based compensation - acquisition of intangible assets	Accumulated deficit	Total
Balance as of December 31, 2017		**19,928**	**924,021**	**-**	**9,972**	**(338,219)**	**615,702**
Net loss for the year						(229,600)	(229,600)
Other comprehensive loss				(51)		-	(51)
Total other comprehensive loss				(51)	-	(229,600)	(229,651)
Transactions with owners:							
Capital increase in connection with issuance of Bonus Shares	3.1*	11			(902)	891	-
Share-based compensation expense	2.5*					2,105	2,105
Total transaction with owners		11	-	-	(902)	2,996	2,105
Balance as of December 31, 2018		**19,939**	**924,021**	**(51)**	**9,070**	**(564,823)**	**388,156**
Net loss for the year						(336,638)	(336,638)
Other comprehensive loss						-	
Total other comprehensive loss				(51)	-	(336,638)	(336,638)
Transactions with owners:						(336,638)	
Capital increase in connection with issuance of Bonus Shares	3.1*	26			(1,197)	1,171	-
Issuance of Matching Shares, net of costs	2.5*	19				-	19
Share-based compensation expense	2.5*					2,131	2,131
Total transaction with owners		45	-	-	(1,197)	3,302	2,150
Balance as of December 31, 2019		**19,984**	**924,021**	**(51)**	**7,873**	**(898,159)**	**53,668**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF CASH FLOWS

PARENT COMPANY

FOR THE YEARS ENDED DECEMBER 31	Note	2019	2018
Operating loss		(335,095)	(231,652)
Reversal of non-cash items:			
Equity-settled share-based compensation expense	2.5*	2,550	2,105
Depreciation, amortization and impliment	2.4, 3.1*, 3.2*	5,010	1,366
Exchange rate adjustments		-	(491)
Change in working capital:			
Change in prepayments, deposits, and other receivables	2.5	5,003	(14,484)
Change in trade payables, accruals and other liabilities	2.6	(2,854)	4,779
Change in intercompany payables		(825)	825
Corporation taxes received	2.7*	5,500	5,500
Interest received (paid)	2.6*	(4,793)	(2,957)
Net cash used in operating activities		**(325,504)**	**(235,009)**
Investing activities			
Purchase of intangible assets	3.1*	(508)	(1,603)
Purchase of property, plant and equipment	3.2*	(2,777)	(743)
Capital increase in subsidiaries			(1,207)
Net cash used in investing activities		**(3,285)**	**(3,553)**
Financing activities			
Proceeds from borrowings	4.2*	62,758	-
Repayment of lease obligations	4.2*	(3,838)	-
Proceeds from issuance of shares	4.2*	19	-
Net cash generated from financing activities		**58,939**	**-**
Net change in cash		**(269,850)**	(238,562)
Effects of changes in exchange rates		(23)	(50)
Cash at the beginning of the year		393,123	631,735
Cash at the end of the year		**123,250**	**393,123**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

SECTION 1

BASIS OF PREPARATION

The financial statements of Orphazyme A/S (the "Parent Company") have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the EU and additional disclosure requirements under the Danish Financial Statements Act.

The Parent Company financial statements of Orphazyme A/S for the year ended December 31, 2019 were approved by the Board of Directors on February 28, 2020 and will be submitted to the shareholders of Orphazyme A/S for approval at the Annual General Meeting to be held on March 26, 2020.

In this section

1.1 CORPORATE INFORMATION

Orphazyme A/S is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. In April 2018, a fully-owned subsidiary, Orphazyme US, Inc., was incorporated in Massachusetts, USA. Orphazyme US, Inc. will directly support the US market to establish closer relationships with the medical, patient, and financial communities.

1.2 SIGNIFICANT ACCOUNTING POLICIES APPLICABLE TO THE PARENT COMPANY

The Parent Company applies the same accounting policies as disclosed in the Group's consolidated financial statements. Therefore, only accounting policies specific to the Parent Company or that differ from the accounting policies applied by the Group are disclosed in these notes to the parent statements. If an accounting policy is not specifically mentioned, the Group accounting policy is applied.

A description of Management's key accounting estimates and judgements as well as new IFRS standards are disclosed in the Group financial statements and also apply to the Parent Company.

The Parent Company financial statements are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Parent Company. Where indicated, amounts are rounded to the nearest thousand, or TDKK.

SECTION 2

NOTES

The notes applicable to the financial statements of the Parent Company are the same as those presented in the Group Consolidated Financial Statements, except for those notes presented in this Section 2.

In this section

2.1 GENERAL AND ADMINISTRATIVE EXPENSES

(TDKK)	2019	2018
External costs	15,507	12,065
Intercompany expenses	9,757	5,510
Employee costs (Note 2.2)	21,366	11,960
Travel and related expenses	1,166	3,893
Pre-commercial activities	1,187	1,625
Depreciation	699	74
Total general and administrative expenses	**49,682**	**35,127**

Intercompany expenses includes general and administrative expenses incurred on behalf of the US subsidiary. Including a write-down of the Investment in Subsidiary in the amount of TDKK 1,207

2.2 EMPLOYEE COSTS

(TDKK)	2019	2018
Employee costs, excluding Executive Management and Board		
Salaries	57,886	36,324
Cash bonus	4,678	2,243
Share-based compensation	1,705	1,006
Pensions	4,926	2,686
Other social security contributions	469	322
Other staff costs	748	881
Total employee costs, excluding Executive Management and Board	**70,412**	**43,462**
Executive Management remuneration		
Salaries	5,189	3,328
Cash bonus	3,313	1,173
Share-based compensation	700	1,139
Pensions	589	372
Other social security contributions	63	4
Other staff costs	101	-
Total Executive Management remuneration	**9,955**	**6,016**
Board and committee fees	2,594	2,584
Travel allowance	294	179
Share-based compensation	145	-
Total Board of Directors remuneration	**3,033**	**2,763**
Total employee costs	**83,400**	**52,241**
Recognized as follows in the Statement of Profit or Loss:		
Research and development expenses	62,034	40,281
General and administrative expenses	21,366	11,960
Total employee costs	**83,400**	**52,241**
Average number of full-time employees	**74**	**44**
Year-end number of full-time employees	**83**	**55**

2.3 INCOME TAXES

The following table presents the total income tax benefit for the years ended December 31, 2019 and 2018:

(TDKK)	2019	2018
Current tax benefit on net loss	75,270	51,722
Tax credit research and development expenses	5,500	5,500
Change in unrecognized deferred tax before tax credit	(74,507)	(51,850)
Permanent differences	(764)	128
Total income tax benefit for the period	**5,500**	**5,500**

The following table presents the reconciliation of the effective tax rate to the statutory corporate income tax rate in Denmark.

(TDKK)	2019	2018
Net loss before tax	(342,138)	(235,100)
Corporate income tax rate in Denmark	22%	22%
Computed income tax benefit	**75,270**	**51,722**
Tax effect of:		
Other non-deductible expenses, including IPO-related costs and share-based compensation	(764)	128
Deferred tax asset not recognized after tax credit	(69,007)	(46,350)
Total income tax benefit for the period	**5,500**	**5,500**

The following table presents the carrying amount of deferred tax in the Statement of Financial Position:

(TDKK)	2019	2018
Tax deductible losses	93,030	61,647
Deferred tax on intangible assets	74,050	35,887
Other temporary differences	759	738
	167,838	98,272
Deferred tax asset not recognized	167,838	98,272
Carrying amount included on statement of financial position	**-**	**-**

2.4 INVESTMENT IN GROUP COMPANIES

§ ACCOUNTING POLICIES

Investments in subsidiaries are measured in the Parent Company financial statements at the lower of cost or recoverable amount.

Any distributed dividends are recognized in the income statmement of the Parent Company.

(TDKK)	2019	2018
Cost at January 1	1,207	-
Additions	-	1,207
Cost end of year December 31	**-**	**1,207**

(TDKK)	2019	2018
Adjustment January 1	-	-
Impairment	(1,207)	-
End of period December 31.	**(1,207)**	**-**

(TDKK)	2019	2018
Carrying amount of investment	**-**	**1,207**

	Registered office	Ownership interest (%)	Share capital	Equity (TUSD)	Net loss (TUSD)
Orphazyme US, Inc.	Delaware, USA	100	1	105	1,550

2.5 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

(TDKK)	2019	2018
Deposits with vendors	295	1,266
Prepayments to vendors	465	633
Leasehold deposit	892	632
Total non-current prepayments and deposits	**1,652**	**2,531**

(TDKK)	2019	2018
Prepayments to vendors	13,174	14,135
Grant income receivable	357	1,237
VAT receivable, net	2,522	1,468
Foreign VAT receivable	1,304	6,116
Other current receivables	1,600	125
Total current prepayments and other receivables	**18,957**	**23,081**

2.6 FINANCIAL ASSETS AND LIABILITIES

(TDKK)	2019	2018
Borrowings (Note 3.5*)	62,824	-
Lease liabilities (Note 3.2*)	12,689	-
Trade payables (Note 3.5*)	1,093	18,090
Accruals	30,080	24,093
Total liabilities measured at amortized cost	**106,686**	**42,183**

(TDKK)	2019	2018
Deferred grant income	95	299
Remuneration to the Board of Directors	1,535	1,836
Lease liability	2,876	-
Payroll and employee related costs	16,277	8,677
Total other current liabilities	**20,785**	**10,812**

2.7 CASH

(TDKK)	2019	2018
DKK	89,154	392,196
EUR	20,083	596
USD	13,972	303
GBP	42	28
Total cash	**123,251**	**393,123**

2019
STATEMENTS AND SIGNATURES

STATEMENT BY THE BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

The Board of Directors and Executive Management have today considered and approved the Annual Report of Orphazyme A/S for the financial year January 1-December 31, 2019.

The consolidated financial statements of the Group and the Parent Company's financial statements have been prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act.

In our opinion, the Group's consolidated financial statements and the Parent Company financial statements provide a fair presentation of the assets, liabilities, and financial position at December 31, 2019 and of the results of the Group's and Parent Company's operations and cash flows for the financial year January 1–December 31, 2019.

In our opinion, Management's Review provides a fair presentation of the development in the Group's operations and financial circumstances, the results of the year, and the overall financial position of the Group as well as a description of the most significant risks and elements of uncertainty facing the Group.

We recommend that the Annual Report be adopted at the Annual General Meeting on March 26, 2020.

BOARD OF DIRECTORS

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Martin Bonde

Anders Hedegaard

Rémi Droller

Martijn Kleijwegt

Sten Verland

Catherine Moukheibir

EXECUTIVE MANAGEMENT

Kim Stratton
Chief Executive Officer

Anders Vadsholt
Chief Financial Officer

INDEPENDENT AUDITORS' REPORT

TO THE SHAREHOLDERS OF ORPHAZYME A/S

OPINION

We have audited the consolidated financial statements and the parent company financial statements of Orphazyme A/S for the financial year January 1 – December 31, 2019, which comprise statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flow, and notes, including accounting policies, for the Group and the Parent Company. The consolidated financial statements and the parent company financial statements are prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act.

In our opinion, the consolidated financial statements and the parent company financial statements give a true and fair view of the financial position of the Group and the Parent Company at December 31, 2019 and of the results of the Group's and the Parent Company's operations and cash flows for the financial year January 1 – December 31, 2019 in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act.

Our opinion is consistent with our long-form audit report to the Audit Committee and the Board of Directors.

BASIS FOR OPINION

We conducted our audit in accordance with International Standards on Auditing (ISAs) and additional requirements applicable in Denmark. Our responsibilities under those standards and requirements are further described in the "Auditor's responsibilities for the audit of the consolidated financial statements and the parent company financial statements" (hereinafter collectively referred to as "the financial statements") section of our report. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

Independence

We are independent of the Group in accordance with the International Ethics Standards Board for Accountants' Code of Ethics for Professional Accountants (IESBA Code) and additional requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these rules and requirements.

To the best of our knowledge, we have not provided any prohibited non-audit services as described in article 5(1) of Regulation (EU) no. 537/2014.

Appointment of auditor

We were initially appointed as auditor of Orphazyme A/S on December 4, 2015 for the financial year July 1 to December 31, 2015. We have been reappointed annually by resolution of the general meeting for a total consecutive period of 5 years up until the financial year 2019.

KEY AUDIT MATTERS

Key audit matters are those matters that, in our professional judgement, were of most significance in our audit of the financial statements for the financial year 2019. These matters were addressed during our audit of the financial statements as a whole and in forming our opinion thereon. We do not provide a separate opinion on these matters. For each matter below, our description of how our audit addressed the matter is provided in that context.

We have fulfilled our responsibilities described in the "Auditor's responsibilities for the audit of the financial statements" section, including in relation to the key audit matters below. Accordingly, our audit included the design and performance of procedures to respond to our assessment of the risks of material misstatement of the financial statements. The results of our audit procedures, including the procedures performed to address the matters below, provide the basis for our audit opinion on the financial statements.

Key audit matters	How our audit addressed the key audit matter
Accrual for costs incurred through Clinical Research Organisations (CROs) *Orphazyme has entered into several clinical research contracts with Clinical Research Organisations (CROs) that perform research and development for the Company.* *Due to the materiality of these arrangements and since they cover multiple periods, recognition of the transactions in the correct period requires Management to make significant estimates.* *We focused on this area due to materiality and because the arrangements and the related accounting treatment of accruals accounting for CRO costs require significant estimation by Management.* *Refer to Note 2.1, 3.4 and 3.5 to the Consolidated Financial Statements.*	*We have evaluated relevant processes including Management's controls to ensure that CRO contracts are recognised and measured appropriately on an ongoing basis.* *We obtained Management's calculations for CRO accruals at year-end and reconciled inputs and key assumptions to underlying terms and conditions in the agreements and relevant internal and external sources – and assessed the accuracy of the accruals for CRO services received – including assessed the accuracy of the accruals in previous periods. Furthermore, the CRO accrual models have been tested for clerical accuracy.* *We assessed whether the disclosures in relation to CRO accruals were appropriate and met the requirements of the accounting standards.*
Funding *In line with the business plan Orphazyme incurred substantial net loss and net operating cash outflows in 2019 and is expected to continue so in 2020.* *Cash and financial assets are monitored on a regular basis by Management and the Board of Directors in assessing current and long-term capital needs.* *Due to uncertainty as to the timing and extend of both cash outflows related to the operations, especially costs associated with clinical trials as well as proceeds from capital increases or other means of funding of Orphazyme, the assessment of capital needs requires management to make significant judgment.* *As at December 31, 2019, the Management and the Board of Directors assessed that addition funding was required in order to fund Orphazyme's operations for the entire 12 months period ending December 31, 2020. Thus, on this basis, measures were taken and on February 7, 2020 Orphazyme completed an offering of 7,032,937 shares in a directed issue and private placement and raised approximately DKK 745 million (see Note 4.7)* *The Management and the Board of Directors assess that the net proceeds from the transaction is sufficient to cover Orphazyme's financing needs until well into 2021.* *We focused on this area due to materiality and because the assessment of capital need requires significant judgment by Management.* *Refer to Note 4.1 and 4.7 to the Consolidated Financial Statements.*	*We obtained Management's financial forecasts for 2020 as approved by the Board of Directors at year-end and reconciled inputs and key assumptions to underlying business plans, trial models developed and similar, as well as other relevant internal and external sources – and assessed the company's ability to fund operations of Orphazyme for at least twelve months from December 31, 2019. In this context, we have also discussed measures taken by management and Board of Directors to secure additional funding to the company. We have tested post balance sheet events, including performed detailed testing of the completed offering of shares and the receipt of approximately DKK 745 million after the balance sheet date.* *We assessed whether the disclosures in relation to the company's capital management procedures and subsequent events disclosures were appropriate and met the requirements of the accounting standards.*

STATEMENT ON THE MANAGEMENT'S REVIEW

Management is responsible for the Management's review.
Our opinion on the financial statements does not cover the Management's review, and we do not express any form of assurance conclusion thereon.

In connection with our audit of the financial statements, our responsibility is to read the Management's review and, in doing so, consider whether the Management's review is materially inconsistent with the financial statements or our knowledge obtained during the audit, or otherwise appears to be materially misstated.

Moreover, it is our responsibility to consider whether the Management's review provides the information required under the Danish Financial Statements Act.

Based on the work we have performed, we conclude that the Management's review is in accordance with the financial statements and has been prepared in accordance with the requirements of the Danish Financial Statements Act. We did not identify any material misstatement of the Management's review.

MANAGEMENT'S RESPONSIBILITIES FOR THE FINANCIAL STATEMENTS

Management is responsible for the preparation of consolidated financial statements and parent company financial statements that give a true and fair view in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act and for such internal control as Management determines is necessary to enable the preparation of financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the financial statements, Management is responsible for assessing the Group's and the Parent Company's ability to continue as a going concern, disclosing, as applicable, matters related to going concern and using the going concern basis of accounting in preparing the financial statements unless Management either intends to liquidate the Group or the Parent Company or to cease operations, or has no realistic alternative but to do so.

AUDITOR'S RESPONSIBILITIES FOR THE AUDIT OF THE FINANCIAL STATEMENTS

Our objectives are to obtain reasonable assurance as to whether the financial statements as a whole are free from material misstatement, whether due to fraud or error, and to issue an auditor's report that includes our opinion. Reasonable assurance is a high level of assurance, but is not a guarantee that an audit conducted in accordance with ISAs and additional requirements applicable in Denmark will always detect a material misstatement when it exists. Misstatements can arise from fraud or error and are considered material if, individually or in the aggregate, they could reasonably be expected to influence the economic decisions of users taken on the basis of the financial statements.

As part of an audit conducted in accordance with ISAs and additional requirements applicable in Denmark, we exercise professional judgement and maintain professional scepticism throughout the audit. We also:

- Identify and assess the risks of material misstatement of the financial statements, whether due to fraud or error, design and perform audit procedures responsive to those risks and obtain audit evidence that is sufficient and appropriate to provide a basis for our opinion. The risk of not detecting a material misstatement resulting from fraud is higher than for one resulting from error, as fraud may involve collusion, forgery, intentional omissions, misrepresentations or the override of internal control.
- Obtain an understanding of internal control relevant to the audit in order to design audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Group's and the Parent Company's internal control.
- Evaluate the appropriateness of accounting policies used and the reasonableness of accounting estimates and related disclosures made by Management.
- Conclude on the appropriateness of Management's use of the going concern basis of accounting in preparing the financial statements and, based on the audit evidence obtained, whether a material uncertainty exists related to events or conditions that may cast significant doubt on the Group's and the Parent Company's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusions are based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Group and the Parent Company to cease to continue as a going concern.

- Evaluate the overall presentation, structure and contents of the financial statements, including the note disclosures, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.
- Obtain sufficient appropriate audit evidence regarding the financial information of the entities or business activities within the Group to express an opinion on the consolidated financial statements. We are responsible for the direction, supervision and performance of the group audit. We remain solely responsible for our audit opinion.

We communicate with those charged with governance regarding, among other matters, the planned scope and timing of the audit and significant audit findings, including any significant deficiencies in internal control that we identify during our audit.

We also provide those charged with governance with a statement that we have complied with relevant ethical requirements regarding independence, and to communicate with them all relationships and other matters that may reasonably be thought to bear on our independence, and where applicable, related safeguards.

From the matters communicated with those charged with governance, we determine those matters that were of most significance in the audit of the consolidated financial statements and the parent company financial statements of the current period and are therefore the key audit matters. We describe these matters in our auditor's report unless law or regulation precludes public disclosure about the matter or when, in extremely rare circumstances, we determine that a matter should not be communicated in our report because the adverse consequences of doing so would reasonably be expected to outweigh the public interest benefits of such communication.

Copenhagen, February 28, 2020
Ernst & Young
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised
Public Accountant
MNE no.: mne33234

Rasmus Bloch Jespersen
State Authorised
Public Accountant
MNE no.: mne35503

Orphazyme A/S (CVR: 32266355)
Ole Maaløes Vej 3
DK-2200 Copenhagen N

Approval at Annual General Meeting (AGM): March 26, 2020
Chairman of AGM: Rikke Schiøtt Petersen, Gorrissen Federspiel Advokatpartnerselskab